“STUDY ON LINEAR DERMATOSES”

Dissertation Submitted in

Partial fulfillment of the University regulations for

MD DEGREE IN

DERMATOLOGY, VENEREOLOGY AND LEPROSY

(BRANCH XX)

MADRAS MEDICAL COLLEGE

THE TAMILNADU DR. M.G.R. MEDICALUNIVERSITY

CHENNAI, INDIA.

APRIL 2015

CERTIFICATE

Certified that this dissertation titled **"STUDY ON LINEAR DERMATOSES "** is a bonafide work done by **Dr. P.SARASWATHY,** Post-graduate student of the Department of Dermatology, Venereology and Leprosy, Madras Medical College, Chennai – 3, during the academic year 2012 – 2015. This work has not previously formed the basis for the award of any degree.

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DECLARATION

I, **Dr.P.SARASWATHY** solemnly declare that this dissertation titled "**STUDY ON LINEAR DERMATOSES**" is a bonafide work done by me at Madras Medical College during 2012-2015 under the guidance and supervision of **Prof. K.MANOHARAN, M.D., D.D.,** Professor and head department of Dermatology, Madras Medical College, Chennai-600003.

This dissertation is submitted to The Tamil Nadu Dr. M.G.R. Medical University, Chennai towards partial fulfillment of the rules and regulations for the award of **M.D Degree in Dermatology Venereology and Leprosy (BRANCH – XX)**

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The dissertation entitled **"STUDY ON LINEAR DERMATOSES"** is a bonafide work done by **Dr. P.SARASWATHY** at Department of Dermatology, Venereology and Leprosy, Madras Medical College, Chennai – 3, during the academic year 2012 – 2015 under the guidance of Professor **Dr.U.R.DHANALAKSHMI M.D.,D.D,DNB.,** Professor, Department of Dermatology, Madras Medical College, Chennai -3.

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CONTENTS

S.NO.	TITLE	PAGE NO.
1.	INTRODUCTION	1-3
2.	REVIEW OF LITERATURE	4-63
3.	AIMS AND OBJECTIVES	64
4.	MATERIALS AND METHODS	65-66
5.	OBSERVATION AND RESULTS	67-93
6.	DISCUSSION	94-101
7.	CONCLUSION	102-103
8.	BIBLIOGRAPHY	
	ANNEXURES	
	PATIENT CONSENT FORM	
	PROFORMA	
	MASTER CHART	
	ABBREVIATIONS	
	ETHICS COMMITTEE APPROVAL	
	PLAGIARISM APPROVAL	

ABSTRACT

INTRODUCTION

Skin lesions present with innumerable patterns like Discoid, Petaloid, Arcuate, Annular, Polycyclic, Livedo, Reticulate, Target, Stellate, Digitate, Linear, Serpiginous and Whorled. Most of the linear lesions follow the Blaschko's lines. Patients with linear lesions attending the Dermatology Out Patient Department at Rajiv Gandhi Government General Hospital comprise study group.

AIMS AND OBJECTIVES

To study the incidence of linear dermatoses , the age and sex incidence, various types of clinical presentation among the patients, the association, various sites of distribution and clinico histopathological correlation of various linear dermatoses.

METHODS AND METHODOLOGY

Detailed history including family history, Martial Status, History of trauma documented with clinical examination. After counselling and after recording their consent for the test, skin biopsy done along with routine investigations and the results are evaluated.

RESULTS

Among the 90 cases, Lichen striatus was the most common presentation followed by linear epidermal naevus and Linear Lichen planus in our study. Family history of similar leisions in any of these patients. Out of 90 cases, 83 cases showed unilateral distribution and only the

remaining 7 showed bilateral distribution of lesions in a linear pattern. 66 cases, had lesions mainly over the extremities, corresponding to the lines of Blaschko.

CONCLUSION

The Incidence of Linear Dermatoses during the period of AUGUST- 2013 to JULY-2014 is 0.2 % Lichen striatus was found to be more common , female preponderance. Majority of patients showed unilateral distribution more on the extremities. Histopathological correlation shows the importance of histopathology which ultimately changing the management in any given condition. Very few associations were noted.

INTRODUCTION

Skin is a very important and largest organ of the body. It is the only organ which is visible and is in direct contact with the environment.

In the examination of the skin, the morphology of individual lesions, their overall pattern and spatial relationship to each other, and their body site distribution are helpful and provide an easily recognizable clue to a rapid visual diagnosis. Indeed, clinical diagnosis is more precise than laboratory tests in many disorders.

Skin lesions present with innumerable patterns like Discoid, Petaloid, Arcuate, Annular, Polycyclic, Livedo, Reticulate, Target, Stellate, Digitate, Linear, Serpiginous and Whorled.

Among these patterns, linearity is a pattern which attracts the attention of patients and clinicians alike. A single lesion may assume a linear shape or a number of lesions may be arranged in a linear pattern.

The mechanisms or anatomical factors dictating the linearity are of the following groups:

Linear configurations determined by the course of blood vessels

- Thrombophlebitis, Mondor’s disease
- Eczema related to varicose veins
- Temporal arteritis

Linear lesions determined by Lymphatic's or Nerve trunks

- Lymphangitis
- Sporotrichosis
- Leprosy

Linear lesions of developmental origin

- Pigmentary demarcation line, Linea nigra
- Epidermal naevi, Incontinentia pigmenti
- Linear psoriasis, Lichen planus, Lichen striatus

Linear lesions following Dermatomal pattern

- Herpes zoster
- Zosteriform nevus
- Darier's disease
- Zosteriform cutaneous metastases

Linear lesion due to stretching of skin

- Striae

Linear lesions caused by External factors like

- Plants, Allergens, Chemicals,

  Thermal and Physical factors

  (Includes Koebner's phenomenon).

Linear configurations due to other determinants

- Linear scleroderma (limb, central forehead)
- Senear–Caroridge (on hands in psoriasis),
- Dermatomyositis (dorsum of fingers; Gottron sign),
- Flagellate pigmentation due to bleomycin

Most of the linear lesions follow the Blaschko's lines. Patients with linear lesions attending the Dermatology Out Patient Department at Rajiv Gandhi Government General Hospital comprise my study group.

REVIEW OF LITERATURE

Lesions in Blaschko’s line and mosaicism

Genetic mosaicism classically appears as Blaschko's lines inthe skin. The pattern may vary according to timing of mosaicism and cell type of involved in mosaicism. This pattern helps to consider what the differential diagnosis would be if the lesions were generalized. If the gonadal mosaicism also present, offspring may present with generalized condition.

INTRODUCTION

The term mosaicism is used in genetics to describe persons with cells of different genotypes. The skin patterns caused by mosaicism are not random. Dermatologist Dr Alfred Blaschko, from Berlin, in 1901[1] stated that epidermal nevi, and some other conditions now known to be mosaic, follow characteristic lines and whorls on the skin. He defined typical striped patterns on the skin, by transferring drawings from numerous patients on to a statue, it was many years before their significance was understood.

Similar striped patterns in mice heterozygous for certain X-linked coat-color genes was reported by Mary Lyon in 1961. She hypothesized that the stripes reflect two populations of cells, one expressing the paternal X chromosome and one the maternal X chromosome, and,

furthermore, that all women are mosaic with regard to the functional X chromosome.

In 1965 Curth& Warburton [2] applied the Lyon hypothesis to the X-linked disorder Incontinentia pigmenti, which is characterized by lesions following Blaschko's lines, and In 1977 Happle [3] recognized lyonization as the cause of Blaschko's lines in female patients heterozygous for other X-linked skin disorders. Now we know that mosaicism is not only due to lyonization but also to somatic mutation, half-chromatid mutation, chromosomal non-disjunction and chimerism can result in Blaschko's lines. These lines are comprehensively reviewed by Bolognia et al [4], and cutaneous mosaicism reviewed more recently by Paller.

Causes of mosaicism and the corresponding pathogenesis [4]

Half-chromatid mutation

A mistake in DNA polymerization during the first meiotic division of gametogenesis, whereby the wrong base is synthesized at one point, resulting in a mismatched double strand. If this mismatched chromosome is passed on to the next generation, the first time it separates in mitosis it will provide two templates that are not exactly complementary, giving rise to two different lines of daughter cells.

Lyonization

The hypothesis, proposed by Mary Lyon, states that only one X- chromosome is active in each female cell, with the other forming the Barr body. Whether the paternal or maternal X chromosome is inactivated is random, but once the choice has been made it is the same in all daughter cells.

Chromosomal non-disjunction

The failure of chromosomes to separate correctly during either meiosis or mitosis, resulting in daughter cells with aberrations of chromosome number or structure.

Postzygotic (somatic) mutation

A mutation occurring after fertilization .

Chimerism

Fertilization of one egg by two sperm, or fusion of two zygotes, resulting in an individual composed of two genetically different cell lines.

Because the clones originate from distinct organisms, chimerism differs from true mosaicism.

MOSAIC SKIN CONDITIONS CLASSIFIED ACCORDING TO THE NATURE OF THE GENERALIZED CONDITION

X-linked dominant (lethal in males)	• Incontinentia pigmenti • Goltz syndrome (chondrodysplasia punctata) • Conradi–Hünermann–Happle syndrome • Congenital hemidysplasia with ichthyosiform nevus and limb defects (CHILD) syndrome • MIDAS syndrome • Oral-facial-digital syndrome type I
X-linked recessive- **(carrier females)**	• Hypohidrotic ectodermal dysplasia • Menkes disease • X-linked reticulate pigmentary disorder • Ectodermal dysplasia, hypohidrotic, with immune deficiency

Autosomal dominant (single gene)	• Linear bullous ichthyosiform erythroderma • Linear Palmoplantar verrucous nevus (keratin 16) • Nevus comedonicus • Linear Darier disease • Linear Hailey–Hailey disease • Linear porokeratosis • Linear basal cell nevi • Linear angiofibromas • Segmental neurofibromatosis - type 1 • Segmental leiomyomas
Autosomal dominant (Possible single gene)	• Nevoid telangiectasia • Linear syringomas • Linear trichoepitheliomas • Linear eccrine spiradenomas

Autosomal dominant / polygenic (Multifactorial inflammatory conditions)	• Linear psoriasis • Linear lichen planus • Linear chronic GVHD • Linear lupus erythematosus • Linear fixed drug eruption • Linear lichen nitidus • Lichen striatus (linear eczema) • Adult blaschkitis • Linear morphea • Atrophoderma of Moulin • Segmental vitiligo
Presumed autosomal dominant lethal disorder rescued by mosaicism (never seen in a generalized form)	• Linear epidermal/sebaceous nevus • Epidermal nevus syndromes • Porokeratotic eccrine ostial and dermal duct nevus (PEODDN)

	• Proteus syndrome • Nevus lipomatosis superficialis • Encephalo cranio cutaneous lipomatosis • Oculocerebro cutaneous syndrome • Inflammatory linear verrucous epidermal nevus(ILVEN) • McCune–Albright syndrome • Zosteriform lentiginous nevus • Localized vascular anomalies • Linear fibromatosis
Chromosomal	• Hypomelanosis of Ito • Nevus depigmentosus • Linear and whorled nevoid hypermelanosis (LWNH)
Chimerism	• Segmental hyperpigmentation

Blaschko's lines represents boundaries between populations of mutant and normal cells, and this was first expressed by Douglas Montgomery [5], who studied several cases of extensive linear epidermal nevi. He concluded that 'the streaks may be due to the streams or trend of growth of the tissues and to the adaptation of the embryonic sutures. Montgomery read his paper before the American Dermatological Association in exactly the same month (May 1901) that Blaschko presented his to the Deutschen Dermatologist chenGesellschaft in Breslau. However, Montgomery's remarkable contribution, published in October 1901, has been published in the same year as his German contemporary. The now widely held idea that Blaschko's lines reflect embryonic cell migration, buthis hypothesis has never been proven.

DRAW BACK

Blaschko's reproduced diagram neglected the scalp, mid-face, lateral neck, and genital area. Now the patterns in above areas , as well as the distributionof lesions in the teeth, eyes and oral cavity were established.[6,7] In the degree of whorling on the flanks, direction on the face and shift of the posterior midline , considerable variation can occur.

Cutaneous mosaicism not follow Blaschko's lines always. Although that cell migration determining the pattern, it depend on when the mosaicism occur during the stages of development. Widely dispersed and intimately mixed clone with long lines will occur, when mutation is earlier. The pattern is also influenced by the processes of lyonization,

lateralization and organogenesis and variation depends on which cell type is affected. The classic pattern stated by Blaschko is on disorders of cells ofepidermis, because Blaschko constructed the drawings from lesions of epidermal nevi [8].

Embryonic keratinocyteson moving away from the neural crest to skin by directional proliferation and forms a continuous line. The tortuous patterns is created by a complex interplay between surface remodeling and cell migration . But Melanoblasts, move away by single-cell migration, and cutaneous proliferation occurs antenatally which is explained by the patchy or phylloid pattern in many of the pigmentary disorders.

Derivaties of cutaneous fibroblasts, blood vessels and other mesodermal tissues will takes direct routes, but rarely shows classic Blaschko's lines corresponding to embryonic dermatomes or segments.

PATTERN OF CUTANEOUS MOSAICISM IN RELATED TO TIMING OF MOSAICISM.

Timing of mosaicism	**Shape in skin**	**Laterality**	**Number of lesion**	**Organs affected**
Meiosis/very early blastocyst	Linear	Bilateral	Multiple	Multiorgan
Late blastocyst/ Pre-organogenesis (First trimester)	Linear	Uni- or bilateral	Fewer	Multiorgan
post-organogenesis (First trimester)	Linear or oval patch	Uni-or bilateral	Few or single	Skin only
Second or third trimester	Oval or round patch	Unilateral	Single	Skin only
Postnatal	Round tumor	Unilateral	Single	Skin only

CUTANEOUS MOSAICISM- EXPECTED PATTERN ACCORDING TO TISSUE OR CELL TYPE

Cell involved	Embryonic migration	Pattern expected
Keratinocyte	Directional proliferation following surface forces	Blaschko
Melanoblast	Single cell migration	Block, Blaschko or phylloid
Fat cell		Segmental
Fibroblast		Segmental
Other mesodermal		Segmental
Blood vessel	Within segments/dermatomes	Segmental/dermatomal
Nerve cell	Along future dermatomes	Dermatomal

THE DIFFERENT PATTERNS OF MOSAICISM [9]

TYPE 1: LINES OF BLASCHKO

Fountain like pattern - back.

S-figure - lateral and ventral aspect of trunk.

Spiral - scalp

These lines reflect the dorsoventral outgrowth of embryonic cells from the neural crest. Their proliferation interfere with the longitudinal growth and increasing flexion of the embryo, resulting in a characteristic arrangement

Head and neck- variable pattern, tent to intersect at an angle of 90˚

TYPE 1.a - narrow bands (e.g.: Incontinentia pigmenti).

TYPE 1.b - broad bands (e.g.: McCune- Albright syndrome).

TYPE 2: CHECKBOARD PATTERN

Flag like area with a sharp midline separation (distributed in a random way and not alternating regularly).

E.g.: speckled lentiginous naevus, Becker nevus

Pattern of patchy hairiness as noted in women heterozygous for

X-linked hypertrichosis

TYPE 3: PHYLLOID PATTERN

Leaf- like patches and oblong macules (midline separation is not always present)

E.g.: Phylloid hypomelanosis (neurocutaneous syndrome)

Phylloid pattern of hyperpigmentation

TYPE4: LARGE PATCHES WITHOUT MIDLINE SEPERATION

E.g.: Congenital giant melanocytic nevi,

with or without neurological Involvement (clonal origin)

Acquired melanocytic nevi

TYPE 5: LATERALIZATION PATTERN

E.g.: CHILD syndrome (X- linked dominant, male lethal-rait)

CHILD nevus-one half of the body, with a sharp midline demarcation.

X-inactivation coincides with the origin of a clone of organizer cells controlling a large developmental field.

TERM ZOSTERIFORM NEVI

A zosteriform arrangement corresponds to the system of dermatomes but all nevi are dermatomal but follow the lines of Blaschko.

LINES THAT DO NOT INVOLVE MOSAICISM

Lines of Voigt-boundaries of peripheral cutaneous innervations.

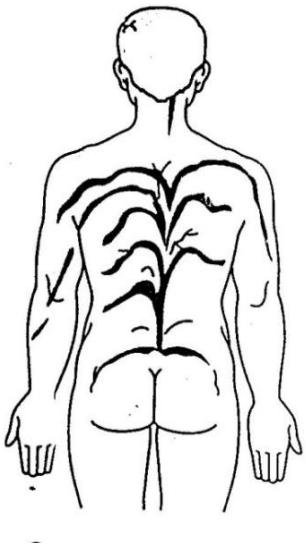
Matsumoto line (also Futcher’s line) pigment demarcation line on arms and legs.

Meridian lines of Acupuncture.

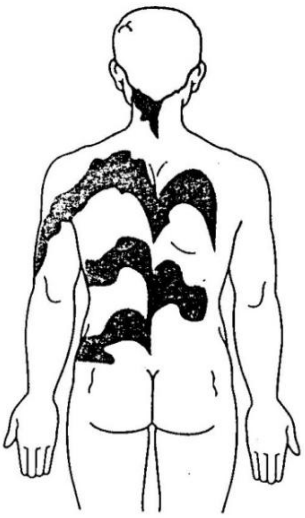
PATTERNS OF CUTANEOUS MOSAICISM

LINES OF BLASHCKO

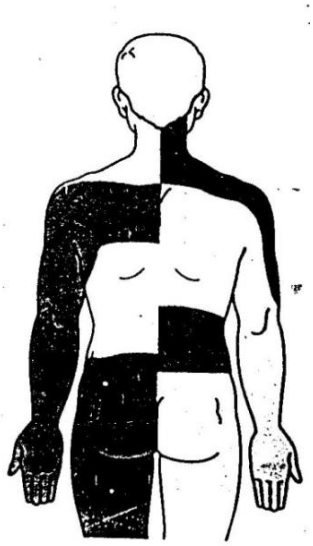
Narrow Band



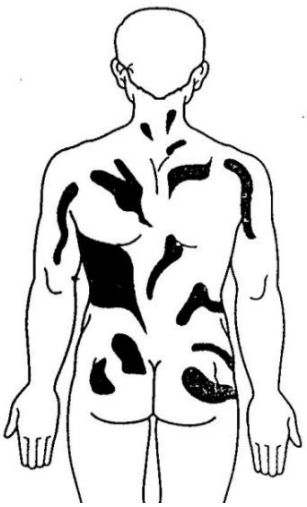
Broad Band



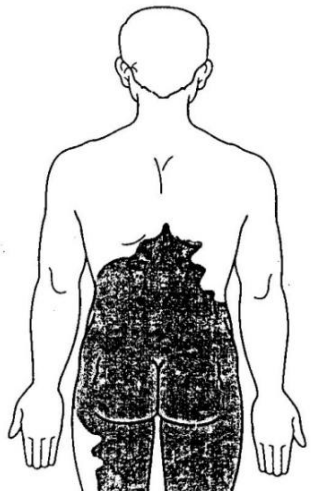
Checker Board



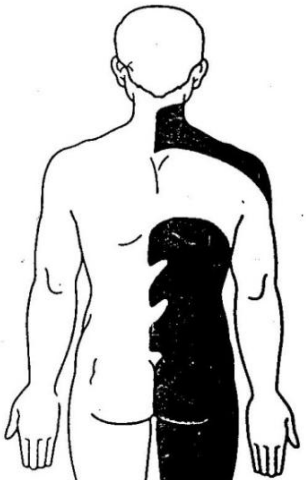
Phylloid



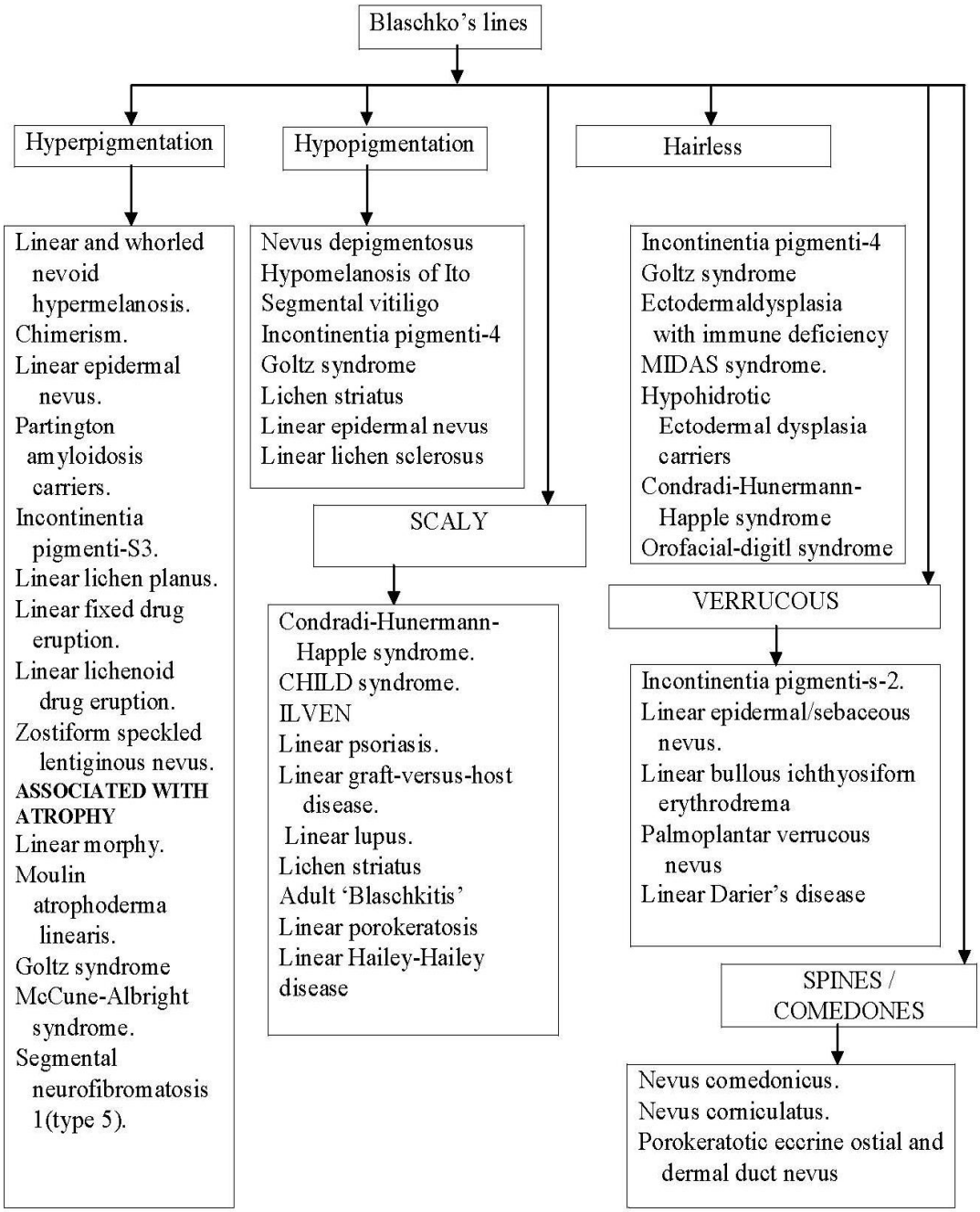
Patchy Pattern without midline separation



Lateralization

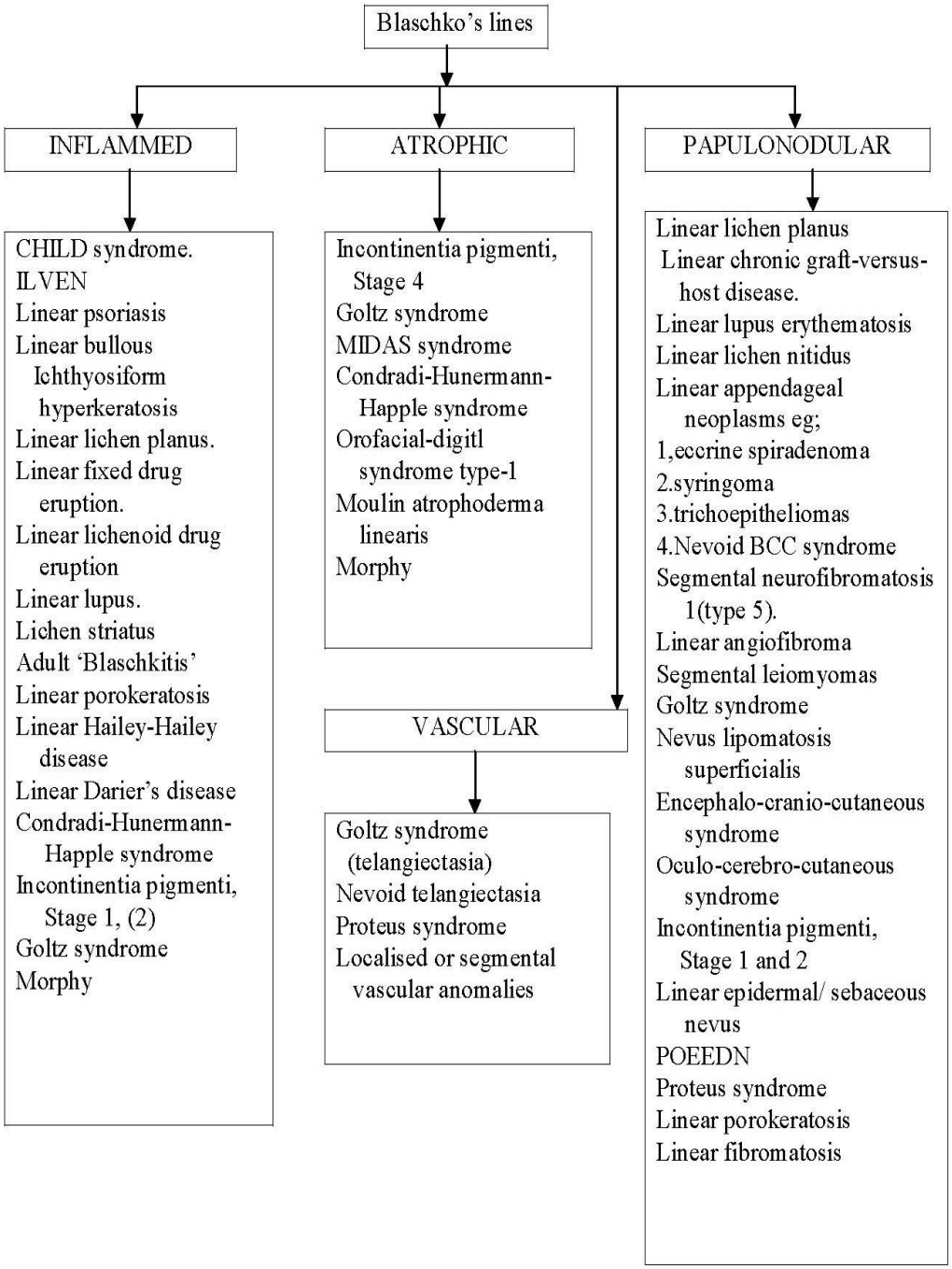


Linear cutaneous lesions that can follow Blaschko's lines[10]

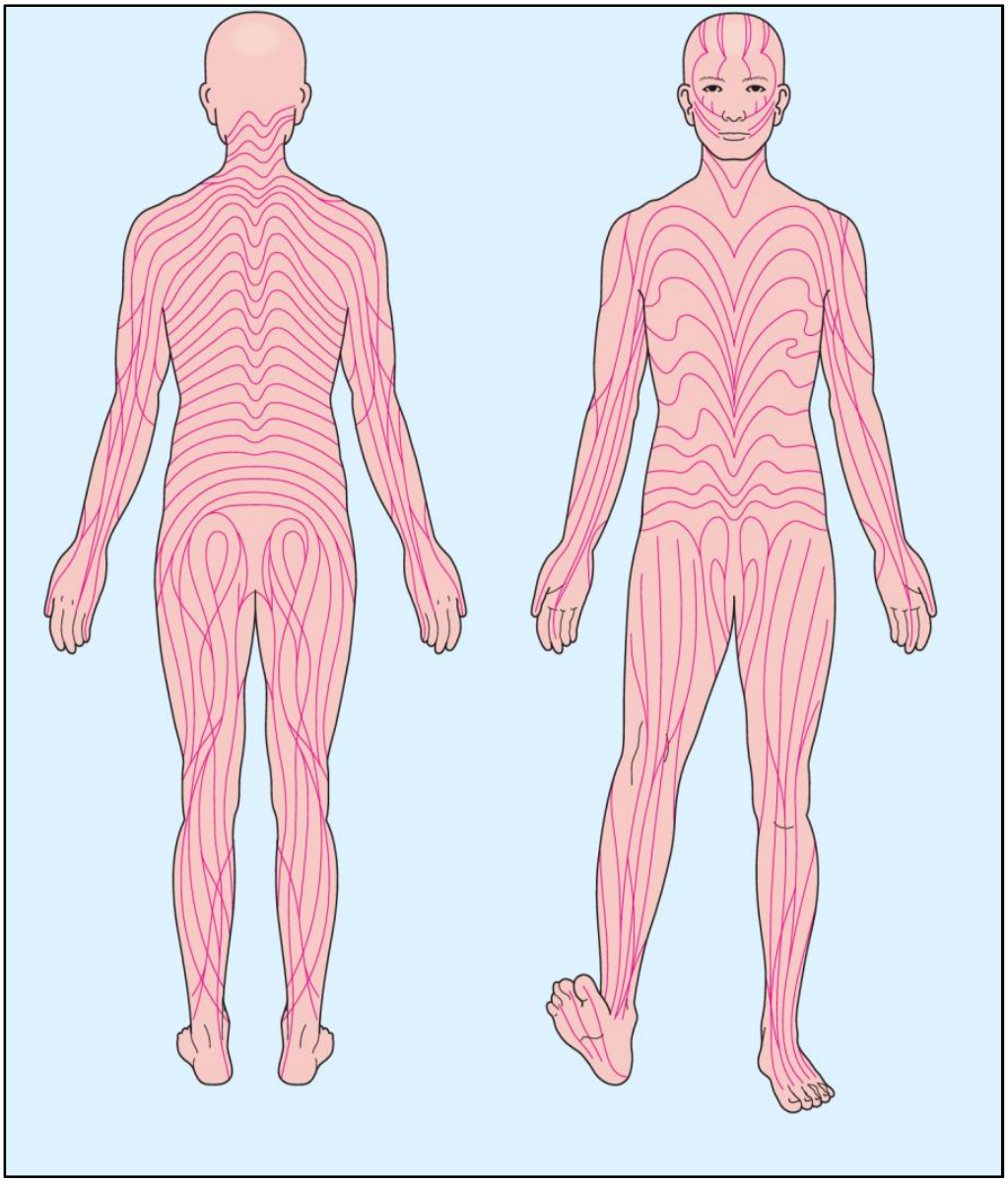


(Conti..)

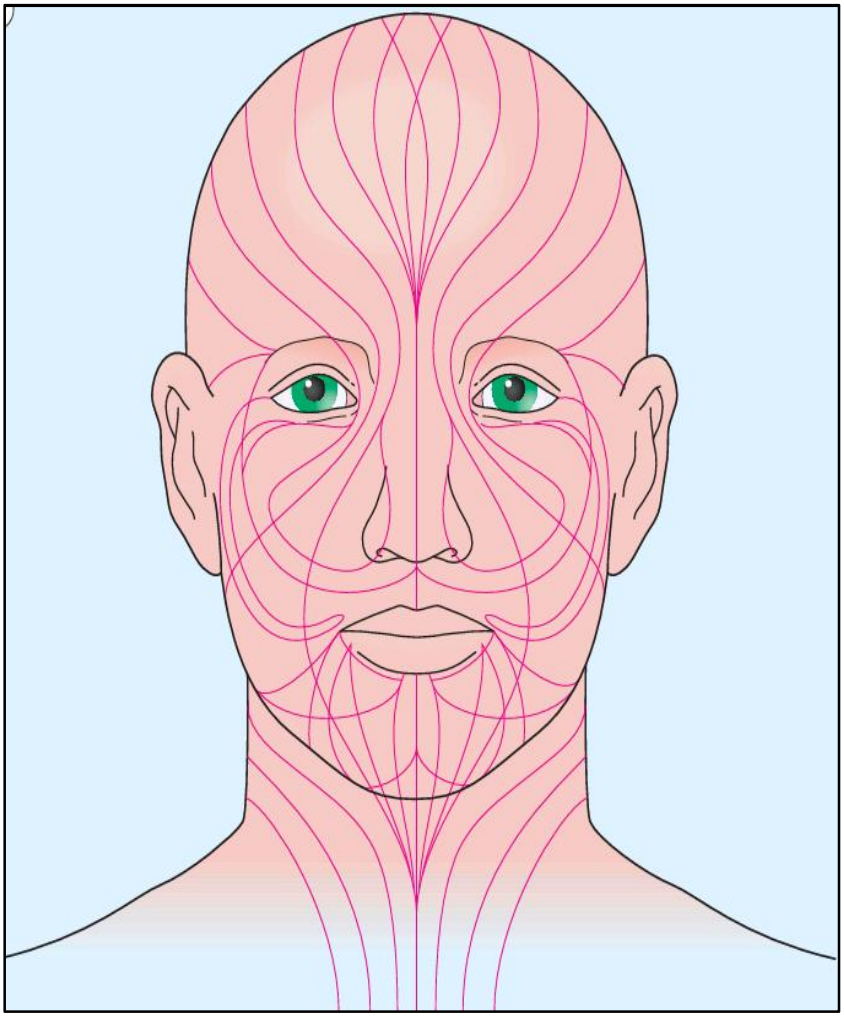
Linear cutaneous lesions that can follow Blaschko's lines
(Conti..)



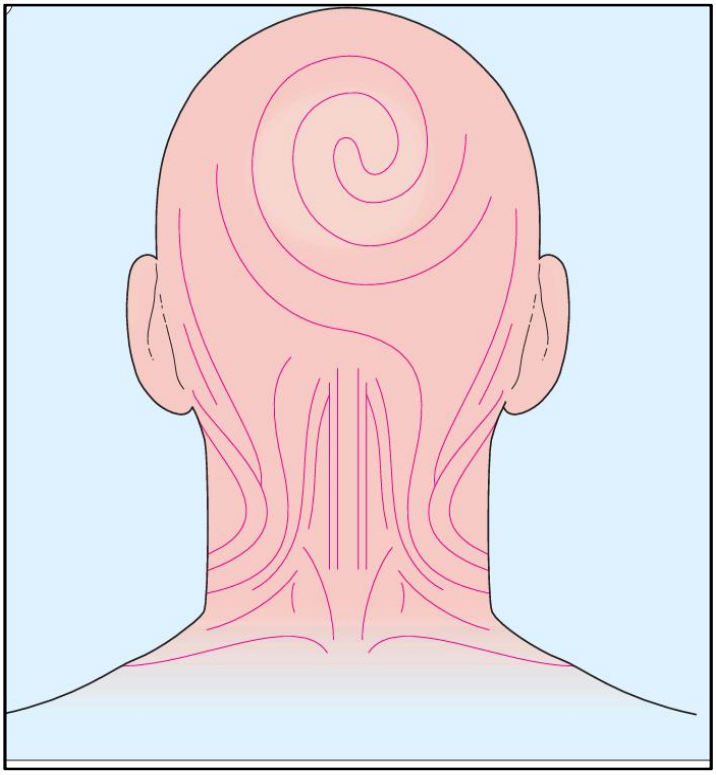
BLASCHKO LINE



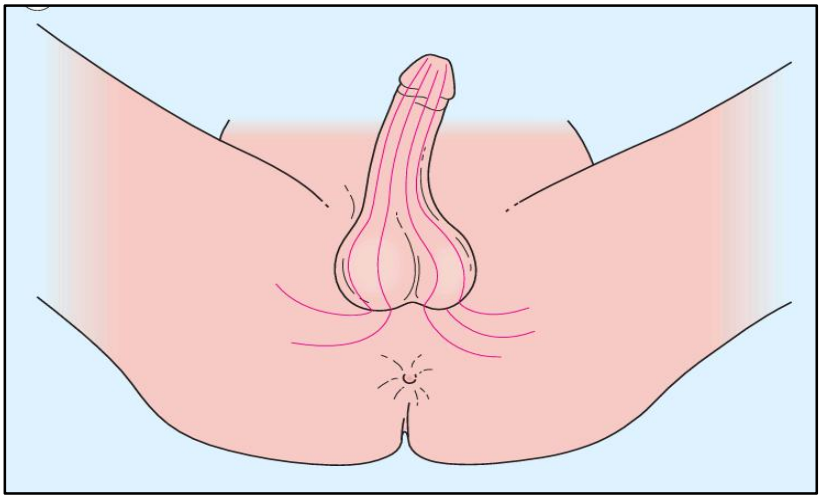
BLASCHKO'S LINES ON THE FACE



BLASCHKO'S LINES ON THE SCALP



BLASCHKO'S LINES ON THE MALE GENITALIA



MOSAICISM IN X-LINKED DOMINANT CONDITIONS

Mosaicism due to random X-chromosome inactivation (lyonization) may produce skin lesions following Blaschko's lines in females heterozygous for X-linked disorders . In females the disorder is compatible with life if recessively inherited and in males it is lethal in utero. In dominant conditions like Incontinentia pigmenti and Goltz syndrome, they arerescued by mosaic pattern . Blaschko's lines in X-linked conditions are mixer of two clones if lyonization also occur simultaneously and are typically narrow and numerous..

Incontinentia Pigmenti

It is a multisystem disorder that may present to neonatologists, neurologists, ophthalmologists, dentists or orthopedic surgeons ,but the diagnosis always depend on the dermatologic findings.

The name is after the pathologic finding of pigmentary incontinence (i.e. dermal melanophages), commonly present during the third stage of the disease. Mosaicism represented by linear skin lesions which is due to lyonization.

History

- 1906-GARROD described first
- 1926-Bloch and Sulzberger contributed their work
- 1900-2 forms IP-1(Sporadic) and IP-2(inherited) due to NEMO gene mutation .

Pathogenesis

The NEMO gene is a subunit of a protein kinase which activates NFκB; TNF-α-induced apoptosis is protected by NFkB. *NEMO* knockout female mice showed patchy granulocyte epidermal infiltration, apoptosis, and then clearance of the cutaneous lesions[11]. Son of IP mother, with severe immunodeficiency and hypohidrotic ectodermal dysplasia is due to milder mutation in NEMO gene.

Clinical feature [12]

Stage-1

Linear erythema and blisters in an apparently normal female new born.

Most common sites of vesiculo bullous lesions are the limbs and scalp, followed by the on the trunk, and rare on the face .

Stage-2

Within days to weeks, these lesions leaves verrucous streaks.

Stage-3

Lines of reticulate hyperpigmentation, the scalloped edges of the lesions consistent with growth of normal keratinocytes into the damaged areas. The inflammatory stage rarely recurs within the pigmented areas during the first few months of life, particularly at the time of febrile illnesses.

During childhood, the hyperpigmented streaks fade usually, sometimes areas of slate-gray pigmentation may persist lifelong.

Stage-4

Linear hypopigmented bands lacking hair and sweating appear on the posterior aspects of the lower extremities, from teenage onwards and it may be the only stigmata of the disease during adulthood. Individual stages may be absent or overlap.

ADDITIONAL MANIFESTATIONS OF INCONTINENTIA PIGMENTI

Ectoderm

- Linear absence of hair and sweat glands
- Missing and conical teeth
- Nail dystrophy
- Nail tumors
- Asymmetric breast development
- Supernumerary nipples

Eye

- Cataracts
- Optic atrophy
- Microphthalmia
- Retinal vascular anomalies
- Pseudoglioma

Central nervous system

- Seizures
- Spastic hemi/di/tetraplegia
- Retardation

Skeleton

- Scoliosis
- Skull anomalies

Respiratory system

- Pulmonary hypertension

Pathology

Stage 1

Inflammatory phase shows eosinophilic spongiosis and scattered dyskeratotic keratinocytes.

Stage 2

Verrucous lesions is acanthotic with hyperkeratosis and foci of dyskeratosis.

Stage 3

Shows pigment incontinence.

Stage 4

Is characterized by a thinned epidermis and dermis devoid of appendages.

Differential diagnosis

Stage 1

Herpes zoster, varicella or herpes simplex viral infections, by the well-being of the child and characteristic patterning of the skin lesions distinguish IP. Eosinophilia can be seen and histologic examination provides confirmation.

Stage2

Linear verrucous epidermal nevus.

Stage 3

Linear and whorled nevoid hypermelanosis(LWNH).

Stage 4

Hypomelanosis of Ito.

Biopsy specimens of the LWNH have predominantly hyperpigmentation in epidermis and absence of dyskeratosis will differentiate it from IP.

GOLTZ SYNDROME

Introduction

Goltz described this rare genetic disorder in 1962, it involves all three embryonic layers

1. Ectoderm derivative- skin & teeth
2. Mesoderm derivative -dermis& bone
3. Endoderm derivative- mucosa of mouth & larynx

Genetics

The mosaicism in skin lesions reflects lyonization. Sporadic cases can be explained by somatic mosaicism and half-chromatid mutations.

Pathogenesis

Mutations in *POCRN* (X chromosome) which encodes a putative O-acyltransferase involved in palmitoylation and secretion of Wnt, a morphogen important in development of ectodermal appendages, dermis, bone and gut.

Clinical features [13]

From birth	-	Streaks of vermiculate dermal atrophy and / or telangiectasias.
Later	-	Hypo and hyperpigmentation as well as outpouchings of fat develop.

Raspberry-like papillomas may appear in any location, they favour the perineal, vulval and perianal regions, as well as the lips and larynx. Bony reduction deformities in the hands particularly a lobster-claw deformity. Skeletal , gastrointestinal, eye, hair and nail abnormalities sometimes occurs.

Pathology [14]

Dermis shows marked reduction in dermal collagen, telangiectasias, varying sizes of fat cells and a decreased number of appendages.

Differential diagnosis

It includes evolving lesions of incontinentia pigmenti, MIDAS syndrome, chondrodysplasia punctata.

CHILD Syndrome

CHILD syndrome is an rare X-linked dominant disorder that, it affects half of the body, with limb reductions, scoliosis, chondrodysplasia punctata, and diffuse erythema with scale. Less complete forms following Blaschko's lines. ILVEN is consider as a limited form of CHILD syndrome. Most cases of CHILD syndrome caused by mutations in a sterol biosynthesis gene, *NSDHL*, on Xq28 [15].

MIDAS Syndrome

MIDAS syndrome is an X-linked dominant disorder characterized by *mi*crophthalmia, *d*ermal *a*plasia, *s*clerocornea and sometimes asssociated with cardiac arrhythmias. It is associated with mutation of*HCCS* on Xp22 which encodes holocytochrome c-type synthase (mitochondria).

Oral-Facial-Digital Syndrome Type I

It is an X-linked dominant disorder characterized by lateral clefts of the jaw and tongue, malformation of the digits, mental retardation, polycystic kidneys, facial milia and patchy alopecia. It is caused by mutations in the *CXORF5* gene at Xp22.3-22.2, a gene widely expressed during development.

Hypohidrotic Ectodermal Dysplasia (Female Carrier)

It is aX-linked recessive disorder, show mild dental anomalies and reduced sweating, which has been demonstrated in a distribution correlating with Blaschko's lines .

Menkes Disease (Female Carrier)

Menkes disease, an X-linked recessive disorder caused by dysfunction of a Cu^{2+}-transporting ATPase (α-polypeptide). Female carriers may show patchy pilitorti and hypopigmentation along Blaschko's lines, attributable to mosaicism for cells with abnormal copper metabolism.

Hypohidrotic Ectodermal Dysplasia with Immune Deficiency

Ectodermal dysplasia, hypohidrotic, with immune deficiency is a recently recognized disorder, in which mutations in the *NEMO* gene produce mild features ofincontinentiapigmenti in women, and ectodermal dysplasia with severe immune deficiency in male offspring and siblings.

MOSAICISM FOR AUTOSOMAL DOMINANT CONDITIONS

Linear autosomal dominant skin disorders are similar in histologically and morphologically to their generalized forms. If there is no generalized disorder coexisting, then linear lesions represents mosaic form of dominant lethal mutation.

It arises either due tohalf-chromatid mutation during gametogenesis or somatic mutation after fertilization. Generally there is no antecedent family history but rare exceptions may be due to unstable permutation .

Epidermolytic hyperkeratosis [16]

Verrucous epidermal nevi also known as ichthyosis hystrix are linear, brown or tan warty lesions often present from birth and distributed along Blaschko's lines. Some have an epidermolytic hyperkeratosis histology identical to the autosomal dominant disorder bullous congenital ichthyosiform erythroderma (BCIE), and they are likewise caused by mutations in keratins 1 or 10.

However as opposed to the generalized disease, in patients with the linear disorder, the mutation is localized to the nevus, while the adjacent uninvolved skin contains the normal gene. Patients with generalized BCIE sometimes inherit their disorder from a parent with the mosaic form, presumably because the parent had gonadal as well as cutaneous mosaicism. It is noteworthy that verrucous epidermal nevi are sometimes associated with cerebral and other systemic manifestations

(epidermal nevus syndrome), but this association is not seen with epidermolytic nevi caused by mutant keratin genes, as they are not expressed in brain tissues.

Unilateral PalmoplantarVerrucous Nevus

It due to mosaicism in keratin 16, Striate PPK present bilaterally and the lesions radiating up to the flexor aspects of the digits. However, Some cases will have unilateral lesions that follows Blaschko's lines. Some consider this as a mosaic form of pachyonychia congenita.

Nevus Comedonicus

Comedonal nevi are present at birth, but become more prominent at puberty under the influence of hormones. They present with numerous keratin-filled pits in a linear distribution, with a sieve-like appearance often on a warty base. Munro observed that they are localized manifestation of generalized disorder Apert syndrome, which is characterized by craniosynostosis, syndactyly, clinodactyly and acne. Munro & Wilkie [17] reported that*FGFR2gene* mutations is responsible for both Apert syndrome and comedonal nevus.

Linear Darier Disease

This is Morphologically and histologically similar to the generalized form. It is due to mutations in the Darier gene *ATP2A.*[18] 'Acantholyticdyskeratotic epidermal nevus' is probably the same disorder as this.

Linear Hailey–Hailey Disease

They are characterized by relapsing linear erosions and crusting, histologically identical to its generalized form due to mutations in the *ATP2C1 gene* confirming type 2 mosaicism[19].

Linear Porokeratosis

This is a linear lesion with of multiple annular plaques similar to typical porokeratosis . It is usually congenital and often lifelong. Though other forms of porokeratosis are with autosomal dominant inheritance, linear lesions reflects mosaic form. Happle suggested that linear porokeratosis represents loss of heterozygosity in a disseminated, which is supported by the occurrence of squamous cell carcinomas [20] only in the linear lesions.

Linear Basal Cell Carcinomas

This is Gorlin syndrome which is characterized by multiple BCCs, broad face, jaw cysts, palmar pits and rib anomalies. This syndrome is due to mutations in the *PTCH* gene on chromosome 9, and represents loss of heterozygosity. [21]..

Segmental Neurofibromatosis- Type 1

These patients have the neurofibromas, café-au-lait macules and / or freckles in a segmental distribution, due to mosaicim for a mutation in the neurofibromin gene. Neurofibromas usually occur in a dermatomal

pattern whereas café-au-lait macules follows Blaschko's lines. Very rarely unstable permutation may result in familial occurrence.

Occasionally, isolate form of plexiform neurofibroma or tibial pseudoarthrosis will represent segmental NF1[22]. Next generation may manifest full-blown condition andthis information should be conveyed to them during genetic counseling.

Linear Angiofibromas

Linear angiofibromas may represent mosaicism of Tuberous sclerosis, but this is not proven. Somatic mutation was demonstrated in a mildly affected parent of an index patient with Tuberous sclerosis, and gonadal mosaicism was detected in unaffected parents who had three affected children affected with Tuberous sclerosis [23].

Nevoid Telangiectasia

It is a unilateral telangiectasia appearing at puberty and often affects women. It may represent mosaic pattern for hereditary telangiectasia.

Other Linear Benign Tumors

Trichoepitheliomas ,Syringomas, and eccrine spiradenomas are inherited ectodermal tumors and sometimes occur in a linear forms . The linear lesions represent genetic mosaicism. Leiomyomas are mesodermal in origin may occur in a segmental pattern occasionally.

LINEAR INFLAMMATORY(MULTI FACTORIAL) DISORDERS

Several inflammatory skin conditions may be distributed in lines of Blaschko. They show a familial susceptibility, suggesting the involvement of atleast one gene in their pathogenesis. They usually appear years after birth, implicating environmental contributions. Several disorders display the Koebner'sphenomenon as well, implying susceptibility factors localized to the skin. The linear patterns of these disorders may reflects mosaicism for a susceptibility mutation.

LINEAR PSORIASIS

Linear psoriasis[24]is very rare and all follows blaschko's lines. There wasthree clinical possibilities to describe this. The first and most common entity is an ILVEN that may resemble psoriasis clinically. Since Woringer's [25] first report in 1936 the psoriasisform nature of some epidermal nevi has been recognized.[26] These lesions often exhibit a characteristic histological picture of areas of hypergranulosis and orthokeratosis alternating with agranulosis and parakeratosis, although features of psoriasis may also be present.

A second entity appears to represent the extension of psoriasis over epidermal nevus by the koebner's phenomenon. Patients may exhibit or develop typical lesions of psoriasis outside the segmental area involved by the nevus [27,28].

A Third entity is ‘linear psoriasis” that does not fit into other types is controversial. Two children were described with extensive lesions following Blaschko’s lines that resembled psoriasis both clinically and histologically has been reported [27].

In both cases there was no pre-existing nevus or any signs of psoriasis in other areas. It is suggested that such presentation may arise due to somatic recombination, giving the linear pattern.

The lesion could be dermatitis epidermal naevi, by their minimal pruritus and their therapeutic response to ultraviolet therapy distinguished them from invasion of a verrucous epidermal nevus by psoriasis as a result of the koebner’s phenomenon but linear psoriasis is most likely confused with ILVEN [26].

LINEAR LICHEN PLANUS

This is a papulosquamous disorder with an insidious in most cases. Familial occurrencedue to genetic predisposition has been reported in monozygotic twins. An increased frequency of lichen planus is noted with HLA-B7, HLA-28, HLA-DR1 and HLA-DR10.

In 1854 Linear LP was first described by Devergie and it accounts for 0.24% - 0.62% of all the patients with lichen planus and it was common in Japan [29].Scattered linear lesions may occur with lichen planus as a result of scratching or due to Koebnerisation. Less commonly, unilateral streaks or bands of LP are seen that are longer and wider than the trauma induced lesions.

In majority of cases, the streaks were formed by polygonal violaceous papules and coalesced plaques showing Wickham's striae. They mainly seen in front of the wrists, lumbar region, ankles and the glans penis.

If it extend to the distal end of digit it may associated with nail dystrophy. Palms, soles and oral mucosa may also be affected.

Multiple linear lesions following the Blaschko's line have been reported in lichen planus. They occur in isolated, long, narrow, linear bands that may extend to whole length of the limb especially in children.[29] Linear LP is more common than linear psoriasis, and it is seen at all ages.

Histopathalogy of linear LP shows compact orthokeratosis, wedge shapedhypergranulosis (clinically Wickham'sstriae), irregular acanthosis and pointed rete ridges, ('saw-toothed' appearance). Liquefactive degeneration of the basal layer, with formation of colloid bodies, melanophages and pigment incontinence. Dense band like inflammatory infiltrate consisting of lymphocytes and histiocytes, closely hugging the lower epidermis will present and the infiltrate also present perivascularly. Lichen planus is an immunologically mediated as evidenced by the immunofluorescence studies [30].

LICHEN STRIATUS

It is a distinctive linear inflammatory papular eruption following blaschko's line, it has to be differentiated from other skin disorders with similar pattern. Variants of this disorder includes

- Blaschkitis,
- Zonal dermatosis,
- Linear neurodermatitis
- Systematized lichenification
- Linear eczema.

Definition

It is a self limiting uncommon linear dermatoses whose etiology is unknown ,with spontaneous regression . It commonly occurs in children from 5-15 years of age , the average age of diagnosis is 3 years.[31,32] sometimes it occurs in adults also. Females are affected more than males and they are affected 2to3 times as frequent as males. [33]

Aetiology

It is unknown though the possibility of an environmental or infective (viral) basis is proposed since some cases cluster in spring and summer. [34,35]

The development of lesion on the blaschko's lines raises the chance of cell-mediated autoimmune reaction to an abnormal clone of

cells. It has been suggested that the distribution of lesions may reflect a post zygotic somatic mutation affecting localized stem cells. Its association with atopy has been reported and about 80% of patients have the family history of atopy [36].

Pathogenesis

It is found that the inflammatory cells reaching epidermis are CD8+ T-lymphocytes , with decreased or increased langerhans cells in the epidermis . These findings gives the possibility of CMI mechanism against keratinocytes can take place during the evolution of the disease. Immunohistochemistry has shown that most of lymphocytes in the epidermis are CD8+T-cells expressing HLA-DR+ antigen on their surface and the lymphocytes in the upper dermis are CD7+. These findings suggest the role of autoimmune mechanism.[37]

Clinical features

They appear as small, pink, lichenoid papules, which are at first discrete, but coalesce into plaques which gives a distinct morphology to this condition[31,32]. They start suddenly then progress in course of a week or more and become a dull red slightly scaly bands.

The width of the band is around 2 mm to 2cm and is usuallyirregular . In course of time these bands may broaden into plaques. The length may vary from few cms to several cms and may extend to the entire length of the limb.

The lesions mostcommonly involves arm or leg and on the neck, but it may rarely occur on the trunk, abdomen, buttocks or the thighs. The lesions are usually unilateral but rarely multiple and bilateral. The lesions are asymptomatic in most of the cases but some may present with itching of moderate to severe degree .

VARIATIONS

- Confluent Verrucous lesions(by johnson)[38]
- Light to yellow grouped papules (byNetherton)[39]
- Flat topped papules (by Frainbell)[40]
- Papules , vesicles and crusting(by Felix pinkus)[41]

Hypopigmentation is prominent in dark skinned persons and it is a useful sign for distinguishing lichen striatus from linear LP. When it extends to distal ends of the digits, it present with nail involvement ranging from fraying to the total nail loss[42] .

Differential diagnosis

It includes linear lesions of lichen planus, lichen nitidus, Epidermal nevus, ILVEN, linear psoriasis and linear lichen simplex chronicus .

Histopathology[43]

Although lichen striatus has been recognized by its variable histologic pictures, chronic lichenoid dermatitis is the most common findings. These changes consists of spongiosis and intra cellular edema

often associated with exocytosis of lymphocytes and focal parakeratosis. Plasma cells and eosinophils are rarely seen. Focal band like distribution in papillary dermis with extension in to the lower portion of the epidermis, vacuolar alternation of the basal layer and necrotic keratinocytes. Dermal findings are superficial perivascular inflammatory infiltrate consisting of lymphohistiocytes . Papillary dermis occasionally contains melanophages.

Less frequently, there are scattered necrotic keratinocytes in the spinous layer as well as subcorneal spongiotic vesicles filled with langerhans cells (EMF Like) [37]. A very distinctive feature is the presence of granulomatous inflammatory infiltrate in the reticular dermis around hair follicles and eccrine glands (Hansens like).

Rarely perforating variant of lichen striatus has been described, which shows transepidermal elimination of clusters of necrotic keratinocytes.

ADULT BLASCHKITIS[44]

These are remitting and relapsing eruption of pruritic inflammatory papules and vesicles. It occurs usually on the trunk in adults. The histology is more eczematous than lichenoid unlike lichen striatus.[45]

It is very difficult to distinguish it from linear grover‘s disease. Taiebetal[46] considered this as adult version of lichen striatus and

proposed the acronym ‘BLAISE’ to cover both. BLAISE should perhaps be regarded as a description rather than a diagnosis and more precise identification are pending till now.

SEGMENTAL VITILIGO

Vitiligo is a multifactorial disorder that occasionally occurs in a segmental distribution. Compared with symmetric vitiligo, the linear type is earlier in onset, less frequently associated with other autoimmune diseases [47], and very rarely spread to other areas of the body. The lesions tend to be patches, broad bands, or blocks, corresponding more to segmental dermatomes rather than along Blaschko's lines, perhaps in keeping with a neuronal pathogenesis. The neuronal abnormality can be mosaic, or alternatively, there can be a clonal susceptibility of melanocytes to neuronal or other influences.

Segmental vitiligo has to be differenciated from nevus depigmentosus. The latter condition is characterized by hypopigmentation rather than depigmentation, and it is present at birth or noted soon thereafter, with a stable size and shape in relation to the growth of the child. The presence since birth of a linear band with complete absence of pigment raises the possibility of mosaicism for piebaldism.

LINEAR MORPHOEA AND PARRY-ROMBERG SYNDROME

It is a localized form of Morphoea which is a characterized by localized sclerosis of the skin of unknown cause. They usually follows Blaschko lines. Provocative factors are trauma , immobilization, BCG vaccination, varicella [48], injections of vitamin K and radiation. Hormonal factors may be the influencing factor since it develop or exacerbate during pregnancy. It appear after measles or Borrelia burgdorferi[49] infection. Familial incidence is also noticed. Frontal type morphoea appears to have a genetic basis.

It may occur in association with phenylketonuria and has occurred in patient on treatment with bromocriptine and pencillamine. Autoimmune aetiology is evidenced by increased incidence of organ-specific auto antibodies in patients and their relatives and it can be rarely associated with idiopathic thrombocytopenic purpura. Females are commonly affected than males (3 times). Peak incidence occurrence is between 20 and 30 years, although 15 % can present in less than 10 years of age.

Linear morphoea is different from other morphoea, with respect to age of onset, distribution, clinical outcome and serology. One distinct aspect is frequent association with high titers of ANA, but biochemical analysis of an individual lesion is indistinguishable from that of classical morphoea.

Linear morphoea presents as erythematous, inflammatory streak, but more frequently it begins as a harmless appearing lesion of plaque-type morphoea that extends longitudinally as a series of plaques that join to form a scar- like band that may severely impair the mobility of the affected limb.

Linear morphoea involves the underlying fascia, muscle and tendons. This results in muscle weakness , shortening of the muscles and fascia, and impaired joint motility when extending over joints. In some patients, involvement is somewhat circular rather than linear and may result in progressive atrophy of the limb similar to the Parry-Romberg variant of facial morphoea.

HISTOPATHOLOGY

i. Early inflammatory stage - found particularly at the violaceous border of enlarging lesions, interstitial lymphoplasmacytic infiltrates present among lighty thickened collagen bundles in the reticular dermis.

ii. Intermediate stage - infiltrates present around eccrine glands, dermis and subcutaneous fat. Large areas of subcutaneous fat are replaced by newly formed fine wavy collagen fibres. Endothelial swelling and edema of vessel wall are also seen.

iii. Late sclerotic stage - inflammatory infiltrate disappears and epidermis is normal. Collagen bundles appear thickened, closely

packed, hypocellular and homogenous. Eccrine glands appear atrophic with few adipocytes surrounding them that leads to high uptake of eccrine gland giving oasis in desert appearance; blood vessels are fibrotic with a narrowed lumen. Elastic fibres are thickened and arranged parallel to epidermis and collagen fibres.

iv. The fascia shows fibrosis and sclerosis. [50]

En-coup -de sabre

This represents linear morphoea of the forehead. It is always unilateral and may extend from the forehead into the frontal scalp. It may start either as a linear streak or a row of coalescing plaques. A paramedian locationis more common than a median location. Like plaque morphoea, it is initially surrounded by a discrete lilac ring and may extends longitudinally reaching the eyebrows, nose and even the cheeks. The waning inflammation leaves a linear, hairless depressed plaque which may be sclerotic or atrophic.

This can also involve the underlying muscles and osseous structures. Rarely, the inflammation and sclerosis progress to involve the meninges and even the brain tissue, resulting in seizures.

Laboratory abnormalities are eosinophilia, elevated ESR and hypergammaglobulinemia. ANA is positive in 67% of patients with linear scleroderma (either speckled or homogenous type).[51] Anti-U1RNP antibodies may present in some cases. Anti single stranded antibody and

Anti-histone antibody may be present which may indicate the aggressiveness of the disease. Rheumatoid factor may also be positive. [52]

Serum level of procollagen type –I carboxy terminal propeptide level is a useful indicator of disease severity with localized morphoea. Increased level of soluble CD23 levels is a new serological indicator of the severity. Scanner using 20MHZ B-mode USG is an non invasive method used to know the thickening and sclerosis of the skin while monitoring the course and treatment of localized scleroderma. [53]

LETHAL DISORDERS RESCUED BY MOSAICISM

Generalized counterpart cannot be recognized for some linear condition. These cases probably represents mutation that would be incompatible with life if it involved all the cells. It affects both men and women and can reflect autosomal dominant or X-linked mutations. There is another possibility that, they could reflect clonal loss of heterozygosity in a normal carrier of a lethal recessive mutation.

EPIDERMAL NAEVI

Epidermal nevus is a developmental malfunction of the epidermis in which is due to excess of keratinocytes, showing abnormal mutation, resulting in a visible lesion with a variety of clinical and histopathological patterns. Epidermal nevus may present either as keratinocyte epidermal nevus, or it may have differentiation towards sweat gland, sebaceous gland or hair follicle.

Verrucous epidermal nevus consist of hyperplasia of the surface epidermis and typically appear as verrucous plaques that coalesce to form well demarcated, skin coloured to brown, papillomatous plaques. Most lesions are present at birth or develop during early infancy. They grow slowly during childhood and they reach a stable size at adolescence.

Lesions may be localized or diffuse. Linear configurations are common, especially on the limbs and may follow skin tension lines or Blaschko's lines. If plaques are minimally elevated and multiple it can be mistaken for Linear and Whorled Nevoid Hypermelanosis.

Epidermal nevi may involve the palms and soles as well as oral mucosa. It may be confused with linear keratoderma, when it occurs over the palm. Additional clinical findings include Wooly hair nevus, straight hair nevus and nail dystrophy.

HISTOPATHOLOGY

Hyperkeratosis, papillomatosis and acanthosis with elongation of rete ridges, epidermolytic hyperkeratosis or focal dyskeratosis. [54]

The salient histological features of epidermolytic hyperkeratosis are

a) Compact hyperkeratosis in the stratum corneum

b) An increased number of irregularity shaped, large keratohyaline granules

c) Perinuclear vacuolization of the cells in the stratum spinosum and in the stratum granulosum.

d) Peripheral to the vacuolization irregular cellular boundaries

Types of Epidermal Nevus (described by Solomon and Esterly)[55]

1. Nevus Unius Lateralis – most common type
2. Icthyosis hysterix
3. Acantholytic epidermal Nevus
4. Nevus Sebaceous
5. Localized linear verrucous nevus
6. Epidermal Nevus of Axilla (Velvety)
7. Mixed type

EPIDERMAL NEVUS SYNDROME *(Solomon syndrome)*

It is a rare condition that refers to the association of epidermal nevi with involvement of other organ system including nervous, skeletal, cardiovascular and urogenital system.[56]

Although most cases occur sporadically Autosomal Dominant transmission has been reported as well. Both males and females are equally affected and age of diagnosis ranges between birth to 40 years.

ETIOLOGY AND PATHOGENESIS

Epidermal nevi are organoid nevi that arise from the pluripotent (stem cells) germinative cells in the basal layer of the embryonic epidermis. Histological variation is common and in different areas of the same lesion different cell components are seen and hence Solomon and Esterly suggested the term *epidermal nevus* to include all the variants.

Epidermal nevus appears to be a form of genetic mosaicism. The concept of AD lethal gene surviving in only mosaic state was proposed by Happle to explain the genetic basis of several syndromes under this condition. Approximately 5–10% of Epidermal nevus may show features of Epidermolytic hyperkeratosis which is a Forme fruste of BCIE (Bullous congenital icthyosiform erythroderma).

CLINICAL FEATURES

Most epidermal nevi are isolated and can occur at any site. Nevus sebaceous is most commonly occur in head and neck area . Lesions are

generally found along Blaschko lines and never cross the midline. Whorled patterns may also seen in the nevi.

In more than 80% of cases the onset is within one year of age. The lesions can extend beyond their original distribution but reaches stability in late adolescence without any further progression.

SYSTEMIC ABNORMALITIES[57]

1. Skeletal-Bone deformities, cysts, hypertrophies and atrophies.
2. Neurological-Mental retardation, seizures (due to hydrocephalus, cerebrovascular malformation and intracranial calcification) and eye defects.
3. Risk of Malignancy – is limited to Nevus Sebaceous.

Visceral malignancy in association includes Wilms tumor, Nephroblastoma, adenocarcinoma of salivary glands , esophagus and stomach.

DIFFERENTIAL DIAGNOSIS

1. Schimmel penning syndrome–sebaceous nevi associated with cerebral abnormalities, coloboma and conjunctival lipodermoid.
2. Nevus comedonicus syndrome – cataracts, skeletal defects and ECG abnormalities.

3. Pigmented Hairy epidermal nevus syndrome- Becker's nevus, Ipsilateral breast hypoplasia and skeletal defects.
4. Proteus syndrome - soft flat epidermal nevi, limb gigantism, skeletal hyperplasia and subcutaneous hamartomas.
5. CHILD syndrome
6. Phakomatosis pigmento keratotica-sebaceous nevus, contralateral lentiginous nevus (speckled).

The importance of this syndrome is screening of patients for associated systemic manifestations. The epidermolytic histological pattern is a mosaicism dueto keratin 1 and 10 mutation. So genetic counseling is required for the epidermolytic type.

NEVUS SEBACEOUS (OF JADASSOHN)

Sebaceous nevi are present in approximately 0.3% of newborns and appear as a waxy to verrucous plaques. Typically, there is a yellow to orange hue that reflects hyperplasia of sebaceous glands. Most commonly over head and neck but also on extremities and trunk . [58] Distribution is along lines of Blaschko, but difficult to appreciate on scalp, face or neck.

In addition to sebaceous nevi, if the patients have ocular, vascular, musculoskeletal and CNS abnormalities, sebaceous nevus syndrome should be diagnosed.

Ocular: Epidermal nevus involving the eyelid (or) conjuntiva, choristomas, cortical blindness, anophthalmia, microphthalmia,

macrophthalmia, corneal opacities, cataracts, Colobomas and lipodermoid tumors of conjunctiva or sclera.[59]

CNS: Seizures, mental retardation and intracranial Hamartomas.

Skeletal: Hypophosphatemic osteomalacia or Rickets.

HISTOPATHOLOGY[59]

Increased number of sebaceous, apocrine ,eccrine glands in the dermis with absent or hypoplastic hair follicles and papillomatosis of the epidermis. Cords of undifferentiated embryonic stage of hair follicle are present in the dermis.

Porokeratotic Eccrine Ostial and Dermal Duct Nevus

This nevus may resembles a comedonal nevus, but it site of occurrence is the palms and soles where pilosebaceous follicles are normally absent. Lesions on adjacent follicle-bearing skin may be verrucous as well as pitted. Two cases have been reported along with linear psoriasis.

PROTEUS SYNDROME[60]

- Body asymmetry, including limb gigantism and macrodactyly
- Linear verrucous epidermal nevi and vascular malformations
- Areas of dermal atrophy with prominent venous patterns
- Palmoplantar cerebriform hyperplasia
- Lipomas

Introduction

Name of the syndrome derived from Greek god of many forms. It is characterized by asymmetrical overgrowth of various tissues. An association with epidermal nevi supports that this is a mosaic disorder.

Genetics

Sporadic in occurrence.

Pathogenesis

The sporadic occurrence and epidermal nevi characteristic of Proteus syndrome suggest a mosaic condition. The development of multiple hamartomas is in keeping with a mutant growth-control gene that is not tissue-specific. The epidermal nevi of Proteus syndrome are similar to those seen individually. Isolated epidermal nevi may represent a limited form of Proteus syndrome, or alternatively, that epidermal overgrowth is controlled at a variety of levels, so that some mutations are more tissue-specific than others.

Recently, germ line mutations in *PTEN*, the gene for Cowden syndrome, have been reported in some patients with Proteus and Proteus-like phenotypes, with loss of heterozygosity in the epidermal nevus.

Clinical features

Common features of Proteus syndrome include localized macrosomia, digital or limb gigantism (especially asymmetric macrodactyly), linear verrucous epidermal nevi, vascular malformations

(particularly port-wine stains), mild dermal hypoplasia (resulting in prominence of subcutaneous veins), cerebriform palmar or plantar hyperplasia, choristomas of the eye, abnormal fat deposition including lipomas and lipoatrophy, and visceral hamartomas.

Pathology

Histopathologic examination of epidermal nevi reveals hyperkeratosis, papillomatosis and acanthosis without additional adnexal differentiation or epidermolytic hyperkeratosis.

INFLAMAMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS (ILVEN)

Altman and Mehregan coined the phrase "ILVEN" to describe a subset of epidermal nevi that were erythematous, inflammed and pruritic. These nevi consists of unique variety of keratinocytic epidermal nevus which has both inflammatory and psoriasiform features. These nevi follows Blaschko's lines.

Etiology includes mosaicism of a dominant mutation, partial expressions of retroposons, clonal dysregulation of growth triggered by HIV infection, absence of involucrin expression in epidermis. [61]

Diagnostic criteria include the following:

- Early age of onset
- Female predominance
- Frequent left lower extremity involvement

- Pruritus
- Classical biopsy finding and
- Lesional persistence with refractoriness

Pathogenesis

Electron microscopy and immunohistochemical studies reveals that keratinocytes differentiation is altered in the parakeratotic areas. Ultrastructurally, keratinocytes have prominent Golgi apparatus and vesicles in their cytoplasm. The intercellular spaces in the upper layers of the epidermis are widened due todeposition of an electron-dense homogeneous material.

The cytoplasm of parakeratotic keratinocytes contains remnants of nucleus and membrane structures and a few lipid droplets. The marginal formation inside the plasma membrane is incomplete, suggesting a deficient keratinization. The majority of the epidermal infiltrating T-lymphocytes are CD8 +.

The lesions occur commonly on the lower extremities, with most common site being the left leg. Although their expression of involucrin is very characteristic, the orthokeratotic epidermis shows almost absence of involucrin. Their pattern differs from psoriasis , since involucrin is expressed prematurely in most of suprabasal keratinocytes in the later. More than 90% of the dermal infiltrate are CD4+ (helper-inducer) T-lymphocytes.

The majority of ILVEN appears during infancy and childhood. Fifty percent are evident by 6 months of life and 75% by 5 years of age. Histological examination of ILVEN demonstrates psoriasiform acanthosis of the epidermis and alternating parakeratosis without a granular layer and orthokeratosis with hypergranulosis. Below the parakeratotic areas, there may be mild exocytosis of lymphocytes and mild spongiosis. The papillary dermis shows mild to moderate perivascular inflammatory infiltrate of lymphohistiocytes [62]

Clinically ILVEN are stable and has response to UV light and topical medications. Treatment of ILVEN is often challenging. The presence of overlapping component of psoriasis will respond to antipsoriatic treatment. Other cases have improved with topical steroids / topical retinoid, oral retinoids and destructive procedures like excision, ablative laser, cryotherapy and dermabrasion. Surgical excisions is an option, but the risk of scarring needs to be considered. Unlike the non-inflammatory epidermal nevi, ILVEN is not associated with CNS defects.

Rarely, there are skeletal abnormalities (reduction deformities) on same side. But these abnormalities may instead represent examples of CHILD syndrome. An alternative name that has been used to describe this association is PEN/ PENCIL (Psoriasiform Epidermal Nevus with Congential Ipsilateral Limb defects).

McCune–Albright Syndrome

Polyostotic fibrous dysplasia is characterized by café-au-lait pigmentation in large blocks, well demarcated at the midline and rarely shows tendency towards Blaschko's lines. The distribution of skin lesions and sporadic occurrence makes it to consider as a mosaic disorder. It is due to mutation in the gene encoding α-subunit of G protein that stimulates adenylatecyclase activity, resulting in over-expression in tissues where cyclic AMP acts as a 'second messenger'.

This explains the patchy endocrine over-activities especially the occurrence of precocious puberty and increased pigmentation due to stimulation of tyrosinase. The distribution of the skin lesions in a large blocks rather than Blaschko's lines, and the involvement of multiple organs shows the occurrence of an early somatic mutation. Large café-au-lait patches that are seen in a healthy individuals may represents a limited expression of McCune–Albright mutation. It has proved more difficult to identify the mutation in skin than in other affected tissues.[63]

Zosteriform Speckled Lentiginous Nevus

It is possible that zosteriform speckled lentiginous nevus and nevus spilus represent mosaicism for potentially lethal mutations. Though it is referred to as zosteriform, these lesions do not have a dermatomal pattern.

Linear Fibromatosis

Multiple fibromas may rarely been described in lines and clusters. Histologically, they show densely packed fibrocytes and histiocytes

Hypomelanosis of Ito (HI)

This multisystem disorder is always presents as hypo pigmentation following Blaschko's lines. Ito from Japan described this in 1952, as a negative image of incontinentia pigmenti . HI affects both sexes, occurs in all races, but seen more commonly in darkly pigmented skin. HI has sporadic occurrence.

Pathogenesis

Chromosomal anomaly is identified in about a one third of patients. The mosaic karyotypic anomalies can affect autosomes or X chromosomes. The phenotype is highly variable, being determined by the particular chromosome defect and the level of mosaicism. Diploid/triploid and 12p tetrasomic mosaicism (eg.Pallister–Killian syndrome) are specific subtypes.

The reason for pigmentary difference between the two cellular clones is not known. All patients have linear dyspigmentation which was explained by disrupted pigment gene by variety of chromosomal abnormalities.

Mosaicism is always present but cannot always be detected. In some cases the clonal abnormality may be too subtle to be detected

cytogenetically, while in others it may be expressed in tissues other than the blood. Skin fibroblasts show chromosomal mosaicism in some patients with normal blood karyotypes; the yield is increased further by studying cultured keratinocytes.[64]

Clinical features

Apart from the cutaneous lesions, the phenotype is highly variable, being determined by the particular chromosomal abnormality present. The hair, CNS, dental, ocular and musculoskeletal system abnormalities are the most frequently reported associations.

Pathology

Lesional skin may appear normal or show reduced numbers of melanocytes.

Nevus Depigmentosus

It is a rare, localized area of depigmented skin, Lesser in 1884 first described it. It is congenital but apparent after birth. There are three clinical types; the commonest is, circumscribed, rounded single lesion, other forms like segmental and systematized forms are very rare, and it may resemble hypomelanosis of Ito clinically.

Lesions commonly occur on trunk. Hairs within the depigmented macules are usually depigmented. Histology may show either reduced or normal melanocytes. A functional defect in melanocytes, with morphological abnormalities of melanosomes, is identified. It remains stable over time and has no other associated findings .[65]

Linear and Whorled Nevoid Hypermelanosis [LWNH]

LWNH is similar to nevus depigmentosus and hypomelanosis of Ito in many aspects , and the term ‘pigmentary mosaicism’ describes all three conditions. The skin findings in LWNH appear at or soon after the birth and are usually fixed. The disorder may be confined to the skin or may associated with the nervous system and cardio vascular abnormalities. LWNH is usually sporadic but Chromosomal mosaicism may also be present. [66] This might be explained by an inherited, structurally X chromosome abnormality due to lyonization. Skin histology shows primarily increased epidermal pigmentation in affected skin as compared to unaffected skin.

LINEAR HYPERPIGMENTATION

Linear hyperpigmentation can result from multiple etiologies, including postinflammatory changes, drug reactions and genetic disorders that result in areas of hypermelanosis along the lines of Blaschko. In the case of pigmentary demarcation lines, the linear hyperpigmentation represents a normal anatomic variant. In the evaluation of a patient with linear hyperpigmentation, an initial step is to determine whether or not the hypermelanosis follows the lines of Blaschko.

PIGMENTARY DEMARCATION LINES

Synonyms

- Futcher's lines
- Voight's lines
- Ito's lines

Introduction

In all races, the dorsal skin surfaces are relatively hyperpigmented compared to the ventral surfaces. In patients with darkly pigmented skin, visible lines of demarcation between dorsal and ventral surfaces are more obvious for example on the anterolateral aspect of the upper arm . These demarcation lines are symmetric and bilateral and are present from infancy and persist throughout adulthood. Additional lines occur on the posteromedial aspect of the thighs, extending from the perineum to the popliteal fossa, as well as the upper chest and in the paraspinal region of the back.

FIVE MAJOR FORMS OF PIGMENTARY DEMARCATION LINES [67]

TYPE- A

A vertical line along the anterolateral portion of upper arm which may extend into the pectoral region.

TYPE-B

A curved line along the posteromedial thigh that may extends from the perineum to the popliteal fossa and even upto to the ankle.

TYPE-C

A vertical or curved hypopigmented band on the mid chest that results in two parallel pigmentary demarcation lines.

TYPE-D

A vertical line in a pre- or paraspinal location.

TYPE-E

Bilateral hypopigmented macules and patches (chest markings) in a zone that runs from the mid third of the clavicle to the periareolar skin.

TYPE-F

V'SHAPED in temple area

TYPE-G

W' SHAPED in temple area

Differential Diagnosis

Postinflammatory hyperpigmentation may occasionally be confused with A, B and D types. However, there is no preceding injury to the skin and the lines are perfectly symmetrical; in addition, these lines present in infancy and are stable in due course. Types C and E pigmentary demarcation lines may be confused with achromic nevi and ash-leaf macules. Linea nigra, the linear hyperpigmentation that extends from the umbilicus to the pubis in pregnant women, is easily distinguished from a pigmentary demarcation line.

AIMS OF STUDY

1. To study the incidence of linear dermatoses at the Dermatology Outpatient Department, Rajiv Gandhi Government General Hospital, Madras Medical College, Chennai
2. To study the age and sex incidence of linear dermatoses.
3. To study various types of clinical presentation among the patients.
4. To study the association of linear dermatoses.
5. To study the various sites of distribution.
6. Clinico histopathological correlation of various linear dermatoses.

MATERIALS AND METHODS

This study includes 90 cases of linear dermatoses. They were assessed clinically and histologically and routine investigations like blood, urine and motion examinations. X-ray tests were done wherever necessary.

Detailed and complete history in all the 90 cases studied was taken. Their address, occupation and socioeconomic status were noted. Special reference regarding the marital status of parents was kept in mind to rule out a genetic basis. Sibling history was taken in all cases to rule out an infectious origin if other siblings were affected by same dermatoses.

Great care was taken to find out associated skin disorders like Alopecia Areata or Nail changes. Special importance was given to rule out koebnerization. Other special findings like Auspitz sign for psoriasis and Wickham's striae for Lichen Planus were noted.

For the cases of Epidermal Nevus Syndrome, opinions of specialty departments like Neurology, Ophthalmology, Oto Rhino Laryngology and Dentistry were sought.

All cases of Lichen Planus, Psoriasis and Lichen Striatus were tested for HBsAg, VDRL and ELISA for HIV. Patients with Linear Morphoea were referred to Rheumatology for further evaluation and for ANA titre.

Complete physical examination was done for each patient with special reference to lymphadenopathy, mucosal changes, Hair changes and Nail changes. Palms and Soles were also examined. After counseling and after recording their consent for the test, skin biopsy done from the advancing edge of the lesions. Biopsy slides were studied with H&E staining.

OBSERVATIONS AND RESULTS

Incidence of Linear Dermatoses

No. of new cases attending our skin OPD, RGGGH, Chennai – per day	120
Total number of new cases attending our OPD during the period of August 2013 to July 2014	44,517
Total cases of Linear dermatoses during this period	90
Incidence of Linear dermatoses	0.2%

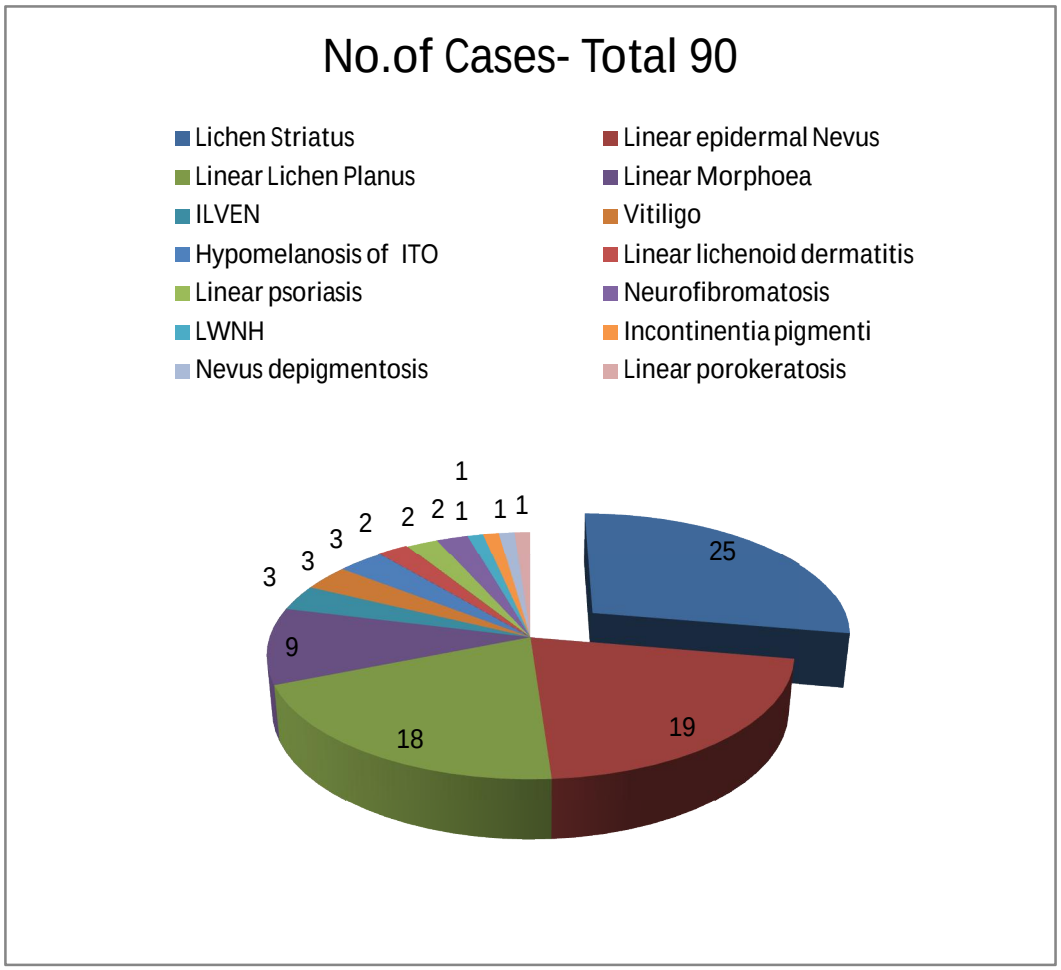
Among the study group of 90 cases, 68 cases were asymptomatic and reported for cosmetic reasons.

Intense itching was the main reason to bring the lichen planus patients and few cases of the lichen striatus patients for treatment.

LIST OF VARIOUS LINEAR DERMATOSES

TABLE 1 (N-90)

Lichen Striatus	25	28%
Linear epidermal Nevus	19	21%
Linear Lichen Planus	18	20%
Linear Morphoea	9	10%
ILVEN	3	4%
Segmental Vitiligo	3	4%
Hypomelanosis of Ito	3	3%
Linear lichenoid dermatitis	2	2%
Linear psoriasis	2	2%
Segmental Neurofibromatosis	2	2%
LWNH	1	1%
Incontinentiapigmenti	1	1%
Nevus depigmentosus	1	1%
Linear porokeratosis	1	1%
Total	90	100%



Among the 90 cases, Lichen striatuswas the most common presentation followed by linear epidermal naevus and Linear Lichen planus in this study. Family history of similar leisions was not present in any of these patients. Out of 90 cases, 83 cases showed unilateral distribution and only the remaining 7 showed bilateraldistribution of lesions in a linear pattern. 66 cases, had lesions mainly over the extremities corresponding to the lines of Blaschko.

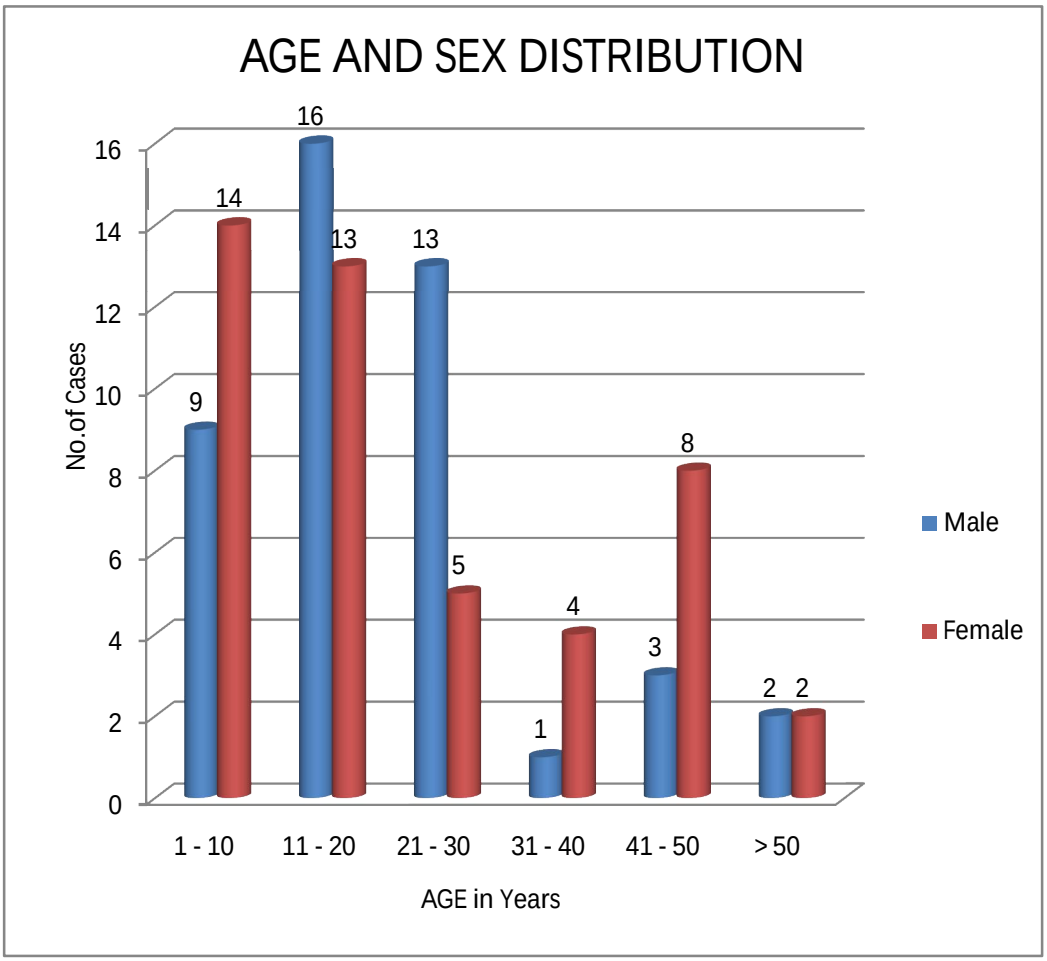
Age and Sex distribution in this study of 90 patients with Linear lesions are shown in Table - 2 (N = 90)

Age (Yrs)	Male	Female	Total
1 - 10	9	14	23
11 - 20	16	13	29
21 - 30	13	5	18
31 - 40	1	4	5
41 - 50	3	8	11
> 50	2	2	4
Total	44	46	90

Total Male cases = 44

Total Female cases = 46

Male: Female Ratio = 10:11



Among the 90 cases, linear lesions was observed most commonly in the age group between 1year to 30 years. Out of ninety patients, 46 were females and 44 were males forming male to female ratio of 10:11.

LICHEN STRIATUS

Among the 90 patients in this group 25 presented with Lichen striatus in the age group between 11 months and 45 years and forming male to female ratio of 11:14.

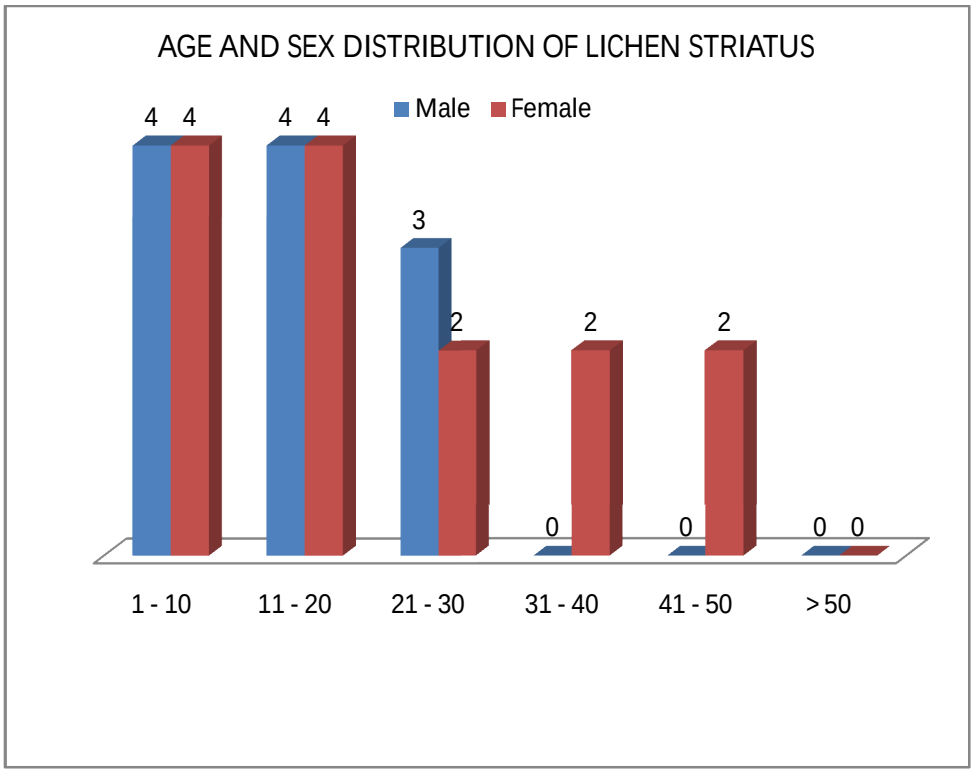
Age and Sex distribution among patients with Lichen striatus are shown in Table - 3 (N = 25)

Age (Yrs)	Male	Female	Total
1 - 10	4	4	8
11 - 20	4	4	8
21 - 30	3	2	5
31 - 40	0	2	2
41 - 50	0	2	2
> 50	0	0	0
Total	11	14	25

Total Male cases = 11

Total Female cases = 14

Male:Female Ratio = 10:14



Sixteen out of 25 patients in this group were in the age group between 1 and 20 years. 20 patients were asymptomatic and mainly came for cosmetic reasons, only 5 patients presented with itching.

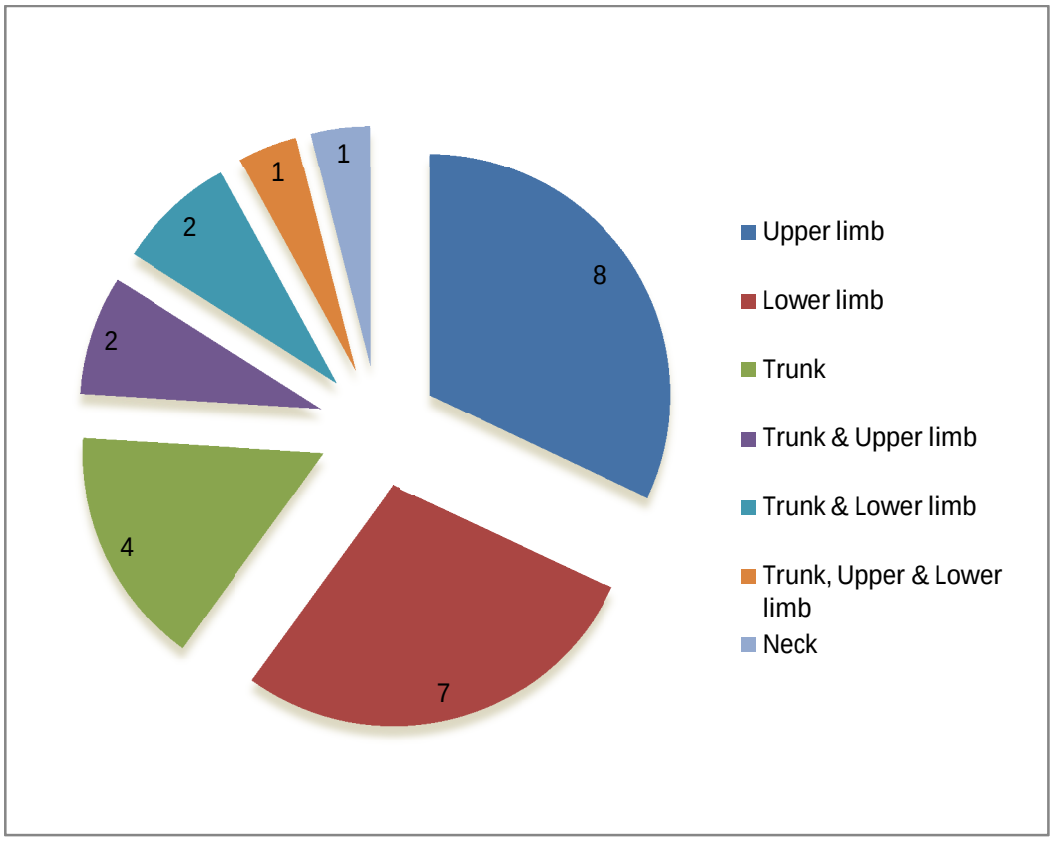
SITES OF DISTRIBUTION OF LICHEN STRIATUS

TABLE -4 (n-25)

Sites of Distribution	No. of Cases
Upper limb	8
Lower limb	7
Trunk	4
Trunk & Upper limb	2
Trunk & Lower limb	2
Trunk, Upper & Lower limb	1
Neck	1
Total	25

Nineteen patients had hypopigmented macules and papules and 4 patients had brown coloured macules and papules and another 2 patients had skin coloured tiny papules. 18 out of 25 cases, had an interrupted linear pattern of Lesions and in the remaining 7 cases Lesions were continuous.

SITES OF DISTRIBUTION OF LICHEN STRIATUS



The Lesions are distributed mainly over the extremities. Multiple site of distribution was noted in 5 patients on same side and the length varying from 7 cms to 30 cms with the mean length of 20 cms.

Most of these Lesions were non scaly and only 7 cases had mild scaling. The duration of the Lesions ranged from 4 months to 4 years, with an average of 12 months.

Among the 25 cases 4 patients had second and third degree consanguineous parentage and none of the other family members of these cases where affected with similar problems.

Skin biopsy was done for 15 cases. Nine cases showed chronic dermatitis picture consisting of mild to moderate acanthosis, mild spongiosis and perivascular lymphohistiocytic infiltrates in the dermis. Two cases showed psoriasiform dermatitis like picture consisting of mild hyperkeratosis, regular acanthosis and sparse inflammatory infiltrates in the upper dermis and blood vessels with spares inflammatory cells seen in the mid dermis.

Three cases showed lichenoid dermatitis like picture consisting of flaky hyperkeratosis mild to moderate acanthosis, indistinct dermo epidermal junction and sub epidermal collections of chronic inflammatory infiltrate. One case showed granulomatous dermatitis picture consisting of flaky hyperkeratosis, keratotic plugging, mild acanthosis, spongiosis, increased pigmentation in basal layer, focal disruption of basal layer with granulomatous inflammatory infiltrate in some area of dermis , around the blood vessels and eccrine glands.

LINEAR LICHEN PLANUS

In this study group 18 patients presented with linear lichen planus. The age group ranged between 7 years and 70 years with an average of 30 years. Out of the 18 cases 11 cases were males and 7 cases were females forming a male: female ratio of 1.5:1.

Age and Sex distribution among patients with Lichen Planus

Table- 5 (n-18)

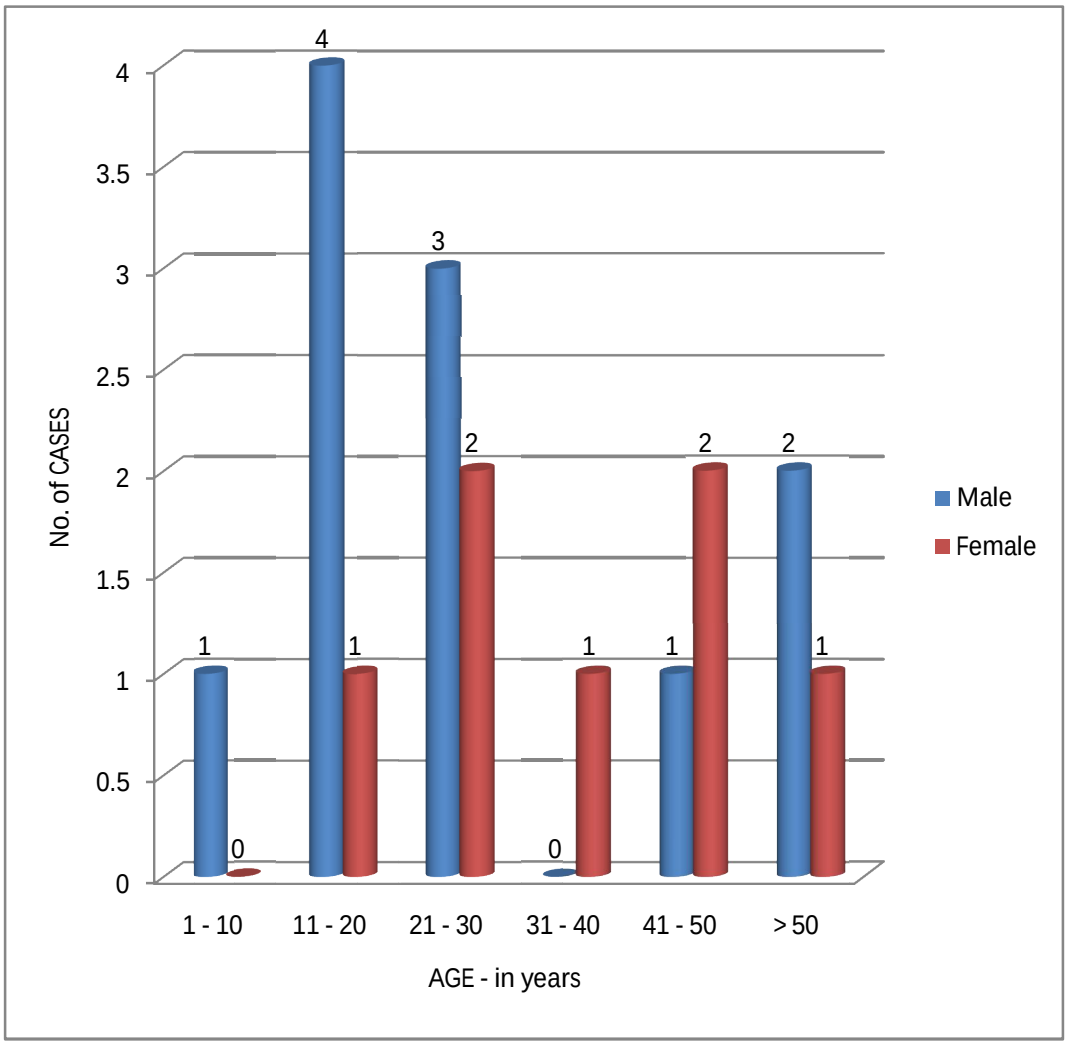
Age (Yrs)	Male	Female	Total
1 - 10	1	0	1
11 - 20	4	1	5
21 - 30	3	2	5
31 - 40	0	1	1
41 - 50	1	2	3
> 50	2	1	3
Total	11	7	18

Total Male cases = 11

Total Female cases = 7

Male: Female Ratio = 1.5:1

AGE AND SEX DISTRIBUTION OF LINEAR LICHEN PLANUS



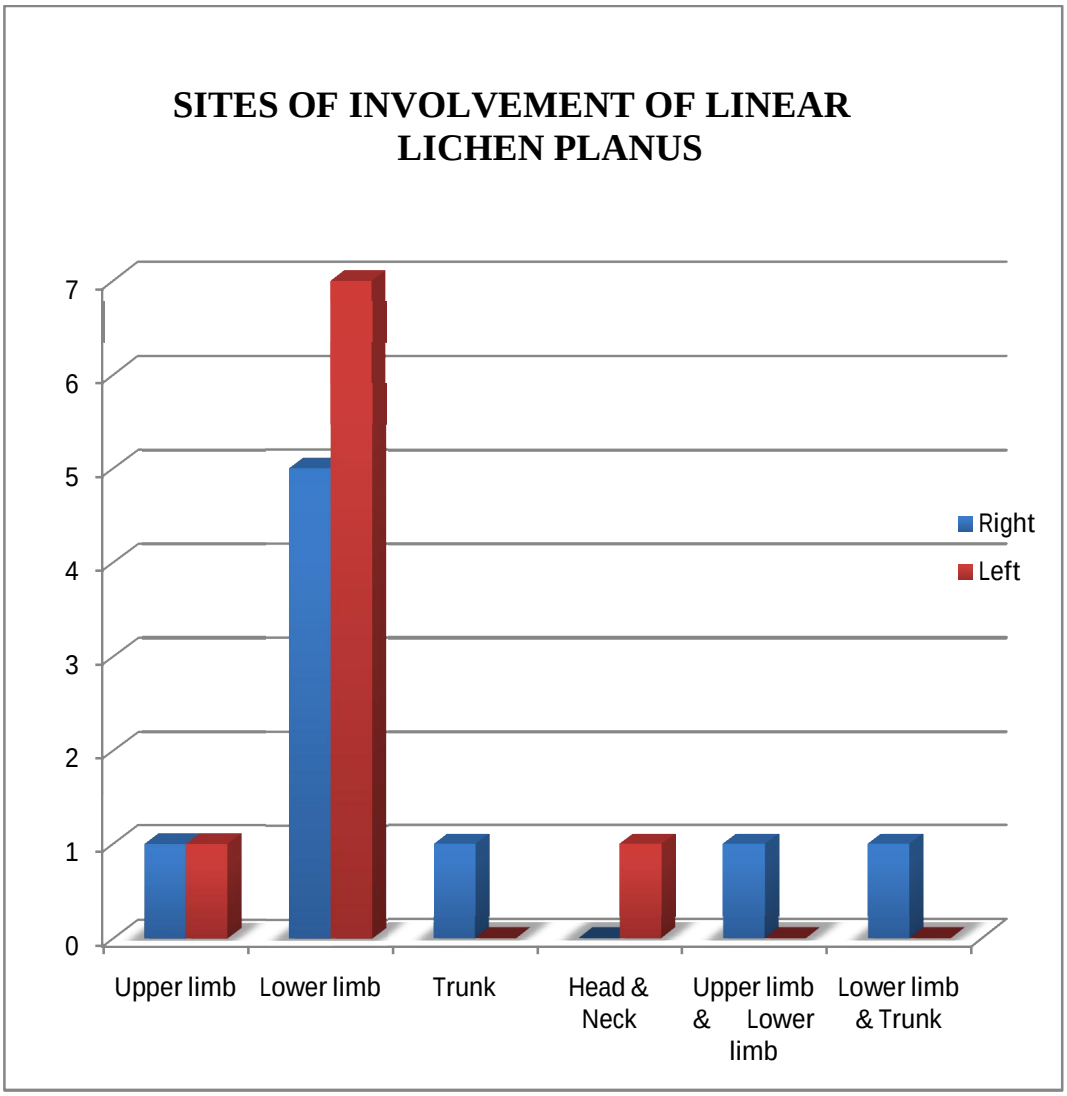
Sites of involvement of Linear Lichen Planus

Table -6 (n-18)

Sites of involvement	Right	Left
Upper limb	1	1
Lower limb	5	7
Trunk	1	0
Head & Neck	0	1
Upper limb & Lower limb	1	0
Lower limb & Trunk	1	0
Total	9	9

Fourteen out of the eighteen cases, presented with itching with duration of symptom ranging from 1 month to 7 years.

Two patients had prior drug intake (NSAID and other unknown drugs) and one had history of hypertenson. Two patients were treated for dermatophytes prior to the onset of lesions.



The lesions were common over the lower extremities especially the left side. Two patient had multiple site involvement, one patient had lesion over the lower limb and trunk . Other patient was presented with the lesions over the right upper and lower limb.

All these patients had hyperpigmented, discrete, flat topped papules and plaques of size varying from 0.5 – 1 cm, with violaceous hue arranged in a linear pattern following Blaschko's lines. The length of the lesions ranged from 5 cms to 50 cms nearly involving the entire limb.

Associated conditions

1. Becker's nevus
2. Pityriasis versicolor
3. Tinea corporis
4. Varicose vein
5. Freckle
6. HbsAg sero positivity
7. Chronic urticaria
8. Androgenic alopecia

None of them showed any mucous membrane involvement, and two patients had nail changes like pterygium and pitting.

Out of eighteen patients biopsy was done for seventeen patients from the recent lesions. Among them 15 specimens showed the classical features of Lichen planus like orthokeratosis, focal hypergranulosis, saw toothed rete ridges, irregular acanthosis, basal cell degeneration, band like lymphohistiocytic infiltrates hugging the epidermis and pigment incontinence. Two specimens showed the classical colloid bodies.

Among the remaining two, one had the features of lichenoid dermatitis like basal cell degeneration, superficial mononuclear cell infiltrate in upper dermis, colloid bodies and pigment incontinence with normal epidermis and other had features of lichen planus pigmentosus.

Linear epidermal nevus

In this study group 19 patients presented with linear epidermal nevus in the age group between 5 years and 55 years and forming male to female ratio of 12 : 7.

Twelve out of 19 patients in the group were in the age group between 1 and 20 years. 15 patients were asymptomatic and mainly came for cosmetic reasons and only 4 patients presented with itching. Sixteen patients had verrucous papules and plaques and 3 patients had skin coloured velvety plaques. 14 out of 19 cases had an interrupted linear pattern of Lesions and in the remaining 5 cases lesions were continuous.

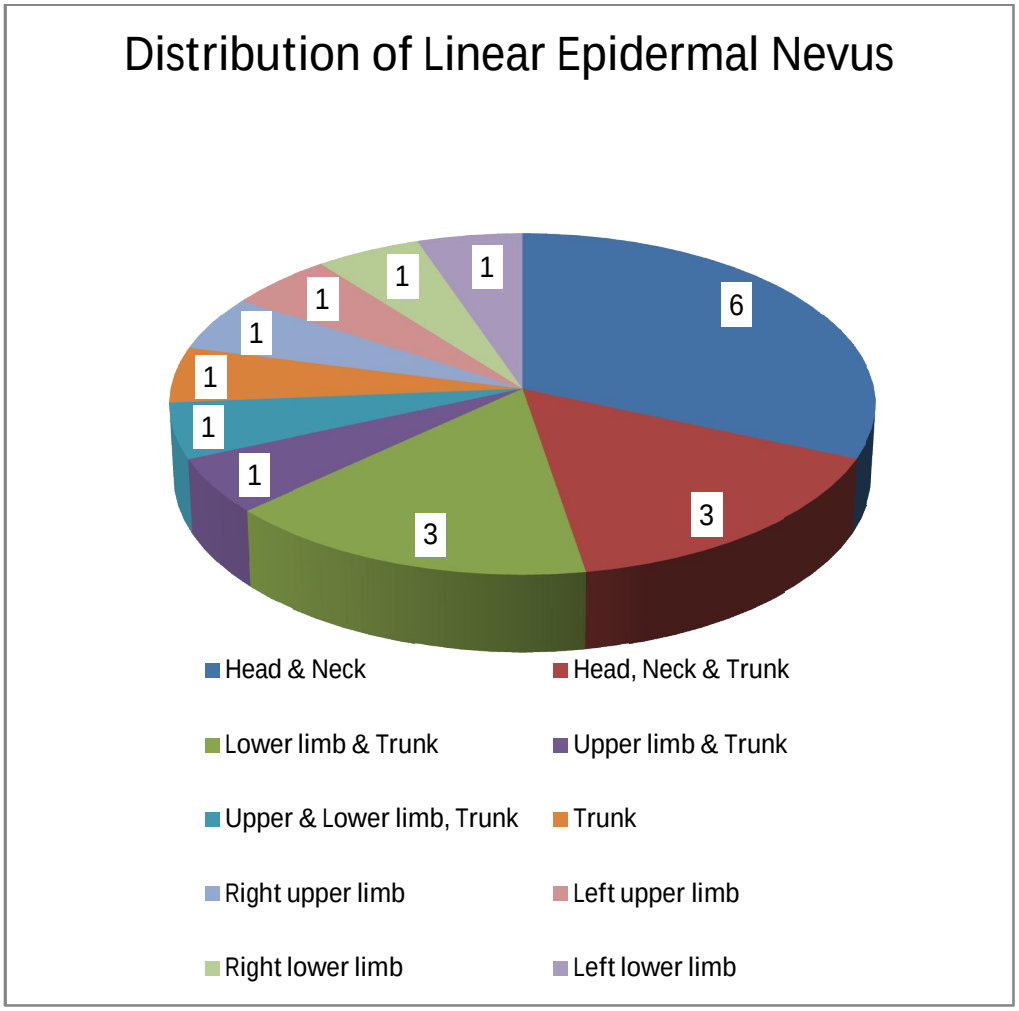
Most of these lesions were brown to black in colour. The duration of the lesions range from 2 years to 25 years with an average of 9 years. Among the 19 patients 2 cases had second and third degree consanguineous parentage and none of the other family members of these cases were affected with similar problems.

Distribution Of Linear Epidermal Verrucous Nevus

Table -7 (n-19)

SITES	No. of cases
Head & Neck	6
Head, Neck & Trunk	3
Lower limb & Trunk	3
Upper limb & Trunk	1
Upper & Lower limb, Trunk	1
Trunk	1
Right upper limb	1
Left upper limb	1
Right lower limb	1
Left lower limb	1
Total	19

The lesions were present mainly over the head and neck. Multiple site involvement is noted in eight patients, and length varying from 6 cms to 50 cms. Eight patient out of nineteen cases had nevi involving trunk implicating systematized form of verrucous epidermal nevus.



Neurological and ophthalmological evaluations were normal for all patients. One patient had unilateral association of gigantism on same side of nevi.

Other associations are

- Lichen simplex chronicus
- TBVC
- Acrochordon

- Acne
- Scabies
- Phrynoderma
- Intertrigo
- One patient had psoriasis over the nevi lesion.

Among the nineteen patients, skin biopsy was done for fourteen cases, which showed the features of Epidermal verrucous nevus like hyperkeratosis, moderate irregular acanthosis, well formed granular layer, increased pigment basal layer, and patchy inflammatory infiltrates in upper dermis.

Two of them showed the features of ILVEN like hyperkeratosis with foci of parakeratosis, moderate acanthosis, elongation and thickening of the rete ridges with a ‘psoriasiform’ appearance, papillomatosis, and slight spongiosis with exocytosis of lymphocytes.

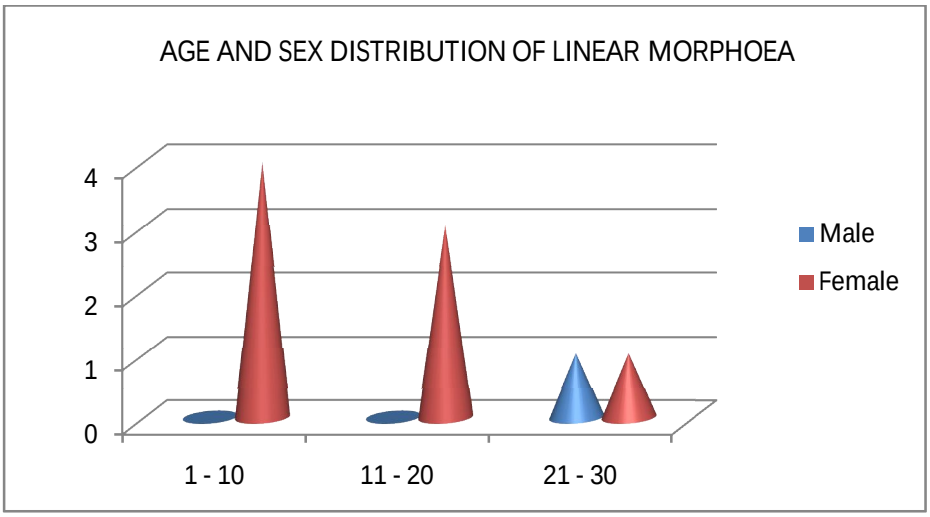
LINEAR MORPHOEA

In this study group 9 patients of linear morphoea was recorded. Two cases were linear pansclerotic morphea, 3 cases were En-coup-de sabre, 3 cases are linear morphoea in extremities and trunk, finally one case was PARRY ROMBERG SYNDROME.

Age and Sex Distribution of Linear Morphoea

Table-8 (n-9)

AGE	Male	Female	Total
1 - 10	0	4	4
11 - 20	0	3	3
21 - 30	1	1	2
Total	1	8	9



The lesions started in early childhood or in adolescence. The duration of lesions varied from 1 year to 4 years. Eight out of the ten cases presented without any specific complaints and the remaining one had pain over the Morphoea plaque. Only one patient had prior history of intramuscular injection over the lesional site.

Only two cases were born of consanguineous marriage (second degree consanguinity). There was no family history of similar illness.

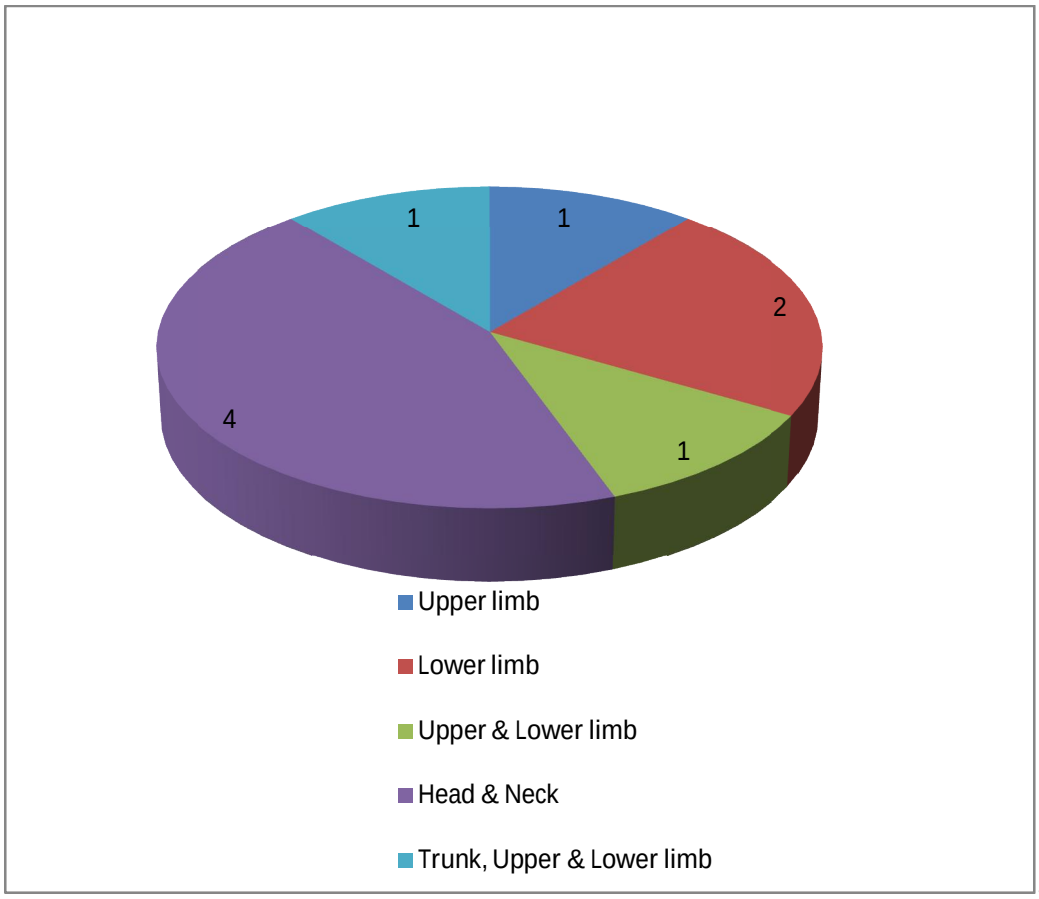
Site of Distribution of Linear Morphoea

Table -9 (n -9)

Sites	No. of cases
Upper limb	1
Lower limb	2
Upper & Lower limb	1
Head & Neck	4
Trunk, Upper & Lower limb	1
Total	9

Four patients had lesions over the head and neck (paramedian), Two patients had lesions involving the lower limbs (left side).Remaining two had lesion over upper limb and lower limb respectively, last patient had lesion over trunk ,upper and lower limbs

SITE OF DISTRIBUTION OF LINEAR MORPHOEA



The lesions were skin coloured to brownish, atrophic, indurated plaques of sizevaring from 3cms to 25cms, arranged in a linear pattern following Blaschko's lines. Three cases of Linear Morphoea were found to be fixed to the underlying structures. All of them had hair loss over the plaques. None of them showed either mucous membrane or nail involvement.

On investigation, eosinophilia was seen in 3 patients and ANA was positive in 2 cases (1/10 and 1/40). X-ray, ECG, EEG and Neurological opinion were sought for all the patients and were found to be normal.

Rheumatologist opinion was obtained for all patients. Biopsy was done for 8 patients. 6 specimens showed the features consistent with early Morphoea like hyperkeratosis, atrophic epidermis, increased pigment basal layer, eosinophilic, edematous collagen bundles in upper dermis and cut section of eccrine duct and arrector pilorum muscle.

Three specimens showed the features consistent with Late Morphoea like atrophic epidermis, collagen bundles appeared homogenous, thickened, and hypo cellular, atrophic eccrine glands narrowed blood vessels and elastic fibres are thickened and arranged parallel to epidermis and collagen fibres.

LINEAR PSORIASIS

In this study two patients were presented linear psoriasis. Both cases presented with hypopigmented scaly plaques of 6 months -2years of duration, over the upper and lower extremities. There is no evidence of any trauma preceding lesions. Both had no similar lesions anywhere else. One of them had pitting over finger nails.

Skin biopsy was done for both the patients which consistent with psoriasis like hyperkeratosis, parakeratosis, absent granular, regular acanthosis, regular elongation of rete ridges, with bulbous thickening of their lower ends, supra papillary thinning of stratum malphigii, irregular dilated tortuous vessels in dermal papillae and perivascular lymphocytes in upper dermis.

SEGMENTAL VITILIGO

In this study group 3 cases presented with segmental vitiligo between the age group 8 years and 21 years.

One patient had lesions over the upper limb, one had lesion over face and other had lesions over the upper limb, neck and face.

The lesions were isolated macules to patches of size 0.5 cm to 10 cms. All the lesions were unilateral in distribution, along the lines of Blaschko. The duration of lesions varied from 3 years to 7 years. The lesions are asymptomatic in all patients. None of them had vitiligo elsewhere are any other associated auto immune disorders. One patient had leukotrichia over the vitiligo patches. Examination of nails and mucosa were found to be normal.

Biopsy was done for 2 cases which showed the features consistent with vitiligo like absence of melanocytes in lesional skin, decrease number of melanocytes in perilesional skin and lymphocytic infiltrates in dermis.

HYPOMELANOSIS OF ITO

In this study 2 females and 1 male patients presented with Hypomelanosis of ITO, between the age group 3 months and 14 years. 2 patients had lesions over the lower limb and abdomen other over the upper limb and abdomen.

The lesions were hypopigmented patches. All the lesions were unilateral in distribution, along the lines of Blaschko. The duration of lesions varied from 2 months to 13 years. The lesions were asymptomatic in all patients. Examination of CNS, Skeletal system and eyes were found to be normal in 2 cases . but one patient is under the treatment for epilepsy with normal neurological investigation.

LINEAR LICHENOID DERMATITIS (LLD)

In this study one female (41 years) and one male patient (24 years) presented with LLD. Both the patients had lesions over the lower limb extensor aspect and they were violaceous plaques. Female patient was with 3 degree consanguinity . There was no history of drug intake prior to the onset of lesions.

SEGMENTAL NEUROFIBROMA

One patient was presented with segmental NF, he is 50 yrs old patient who was under the treatment for pemphigus vulgaris. Lesions were present in right hypochondria of abdomen ,button hole sign was positive. Patient was known diabetic for the past 5 years.

ILVEN

In this study group 3 patients recorded as ILVEN. Age group was between 9 years to 44 years. Two patients had lesions over the lower limb and the other one presented over the trunk.

The lesions were multiple scaly patches and plaques of size 1cm to 5cms.All lesions were unilateral in distribution along the line of blaschko. The duration of the lesions varied from 3 months to 5 years. One patient was diagnosed clinically as lichen striatus, but histopathology showed the features of ILVEN.

INCONTINENTIA PIGMENTI

10years old female patient presented with hypopigmented and hyperpigmented atrophic patches over trunk, extremities and thigh since birth associated with aplasia cutis, right limb hypotonia, limb length discrepency, hypoplastic external genitalia, and cicatricial alopecia over right side of scalp. Histopathology showed hyperkeratosis, keratotic plugging, spongiosis in some areas. Suprabasal cleft present. Increased pigment in basal layer. Pigment incontinence in the upper dermis with inflammatory infiltrate in the upper dermis mainly around the blood vessels.

LINEAR POROKERATOSIS

22 yrs male born out of 3 degree consanguinity, presented with hypopimented lesion in left foot of 5 years duration. It was associated with tinea cruris and longitudinal melanonichia in left thumb. Histopathology showed coronoid lamella, hyperkeratosis, hypogranulosis, acanthosis and lymphocytic infiltrate in upper dermis

Linear Whorled Nevoid Hypermelanosis

14 years male patient presented with multiple linear and whorled hyperpigmented lesion all over the body. Echo,x-ray, EEG was normal. Biopsy showed increase pigmentation in basal layer.

NEVUS DEPIGMENTOSUS

40 years male patient presented with hypopigmented patch in right side of face since birth.

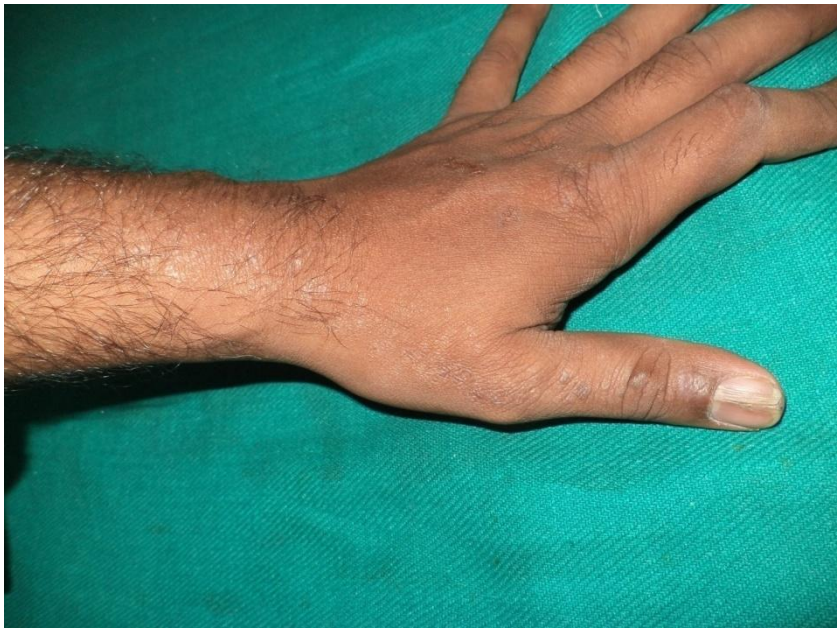
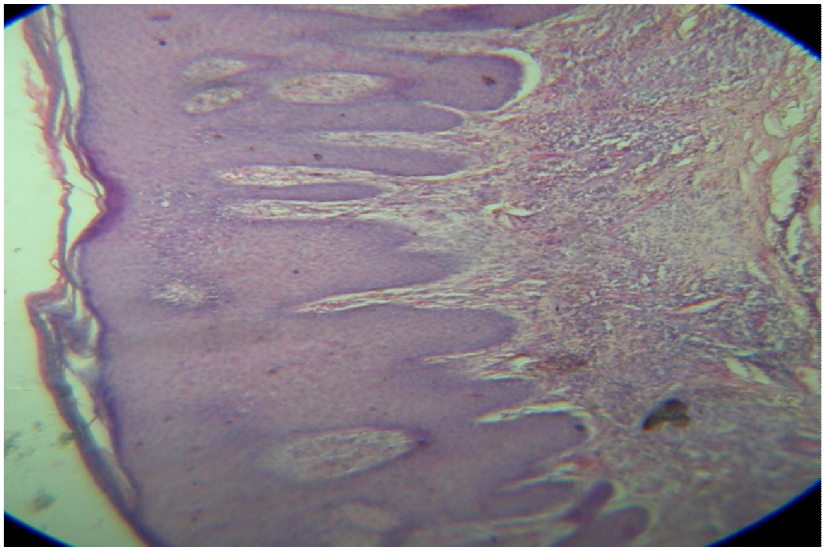
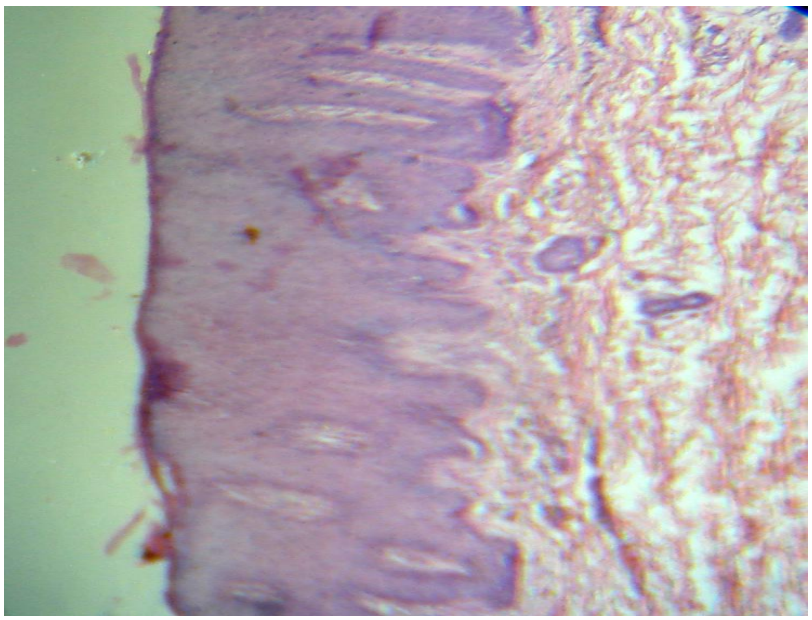


FIG 1. LICHEN STRIATUS



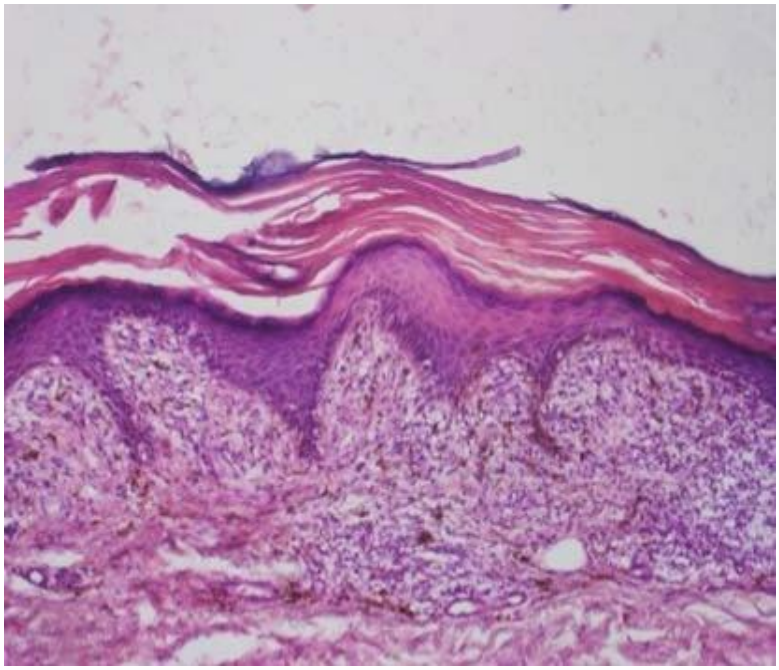
HPE OF LICHEN STRIATUS SHOWING CHRONIC DERMATITIS PICTURE

FIG 2.LICHEN STRIATUS



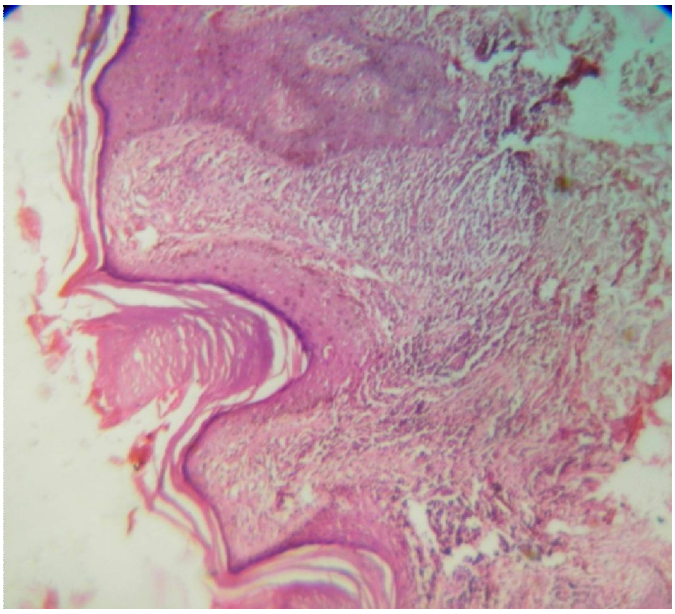
HPE OF LICHEN STRIATUS SHOWING LICHENOID DERMATITIS PICTURE

FIG 3.LINEAR LICHEN PLANUS IN LOWER LIMB IN TWO PATIENTS



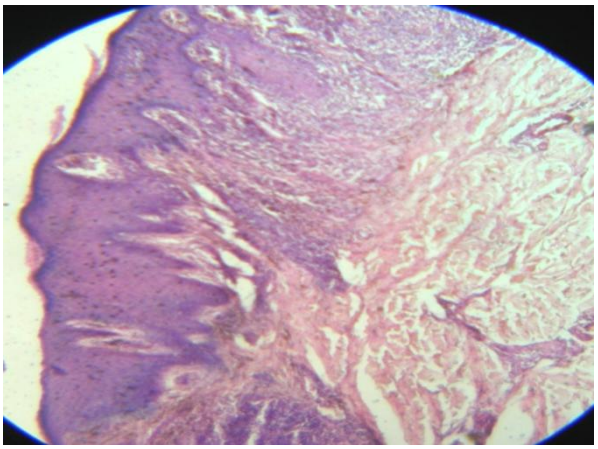
HPE OF LINEAR LICHEN PLANUS SHOWING BASAL CELL DEGENERATION, BAND LIKE INFLAMMATORY INFILTRATES IN THE DERMO-EPIDERMAL JUNCTION

FIG 4.LINEAR LICHEN PLANUS INVOLVING THE LOWER LIMB



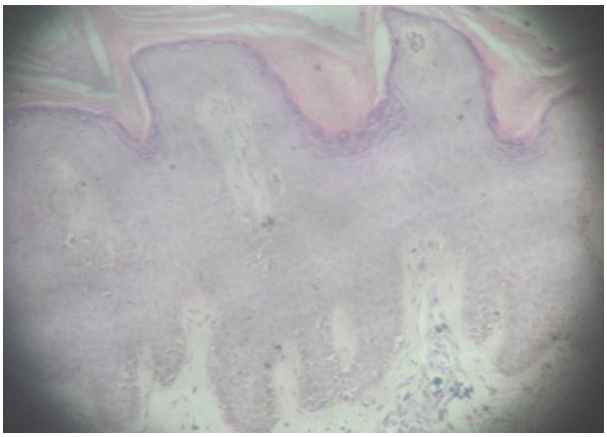
HPE OF LINEAR LICHEN PLANUS HYPERKERATOSIS, BAND LIKE INFILTRATES AND COLLOID BODIES

FIG 5. LINEAR LICHEN PLANUS WITH PTERYGIUM



HPE OF LINEAR LICHENOID DERMATITIS SHOWING BASAL CELL DEGENERATION, MONONUCLEAR CELL INFILTRATES IN UPPER DERMIS AND PIGMENT INCONTINENCE

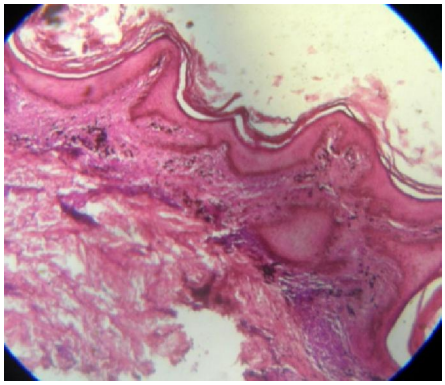
FIG 6.LINEAR VERRUCOUS EPIDERMAL NEVUS



HPE OF LINEAR EPIDERMAL NEVUS SHOWING HYPERKERATOSIS, PAPILLOMATOSIS, ACANTHOSIS , ELONGATION OF RETE RIDGES OF EPIDERMIS AND FEW INFILTRATES IN UPPER DERMIS

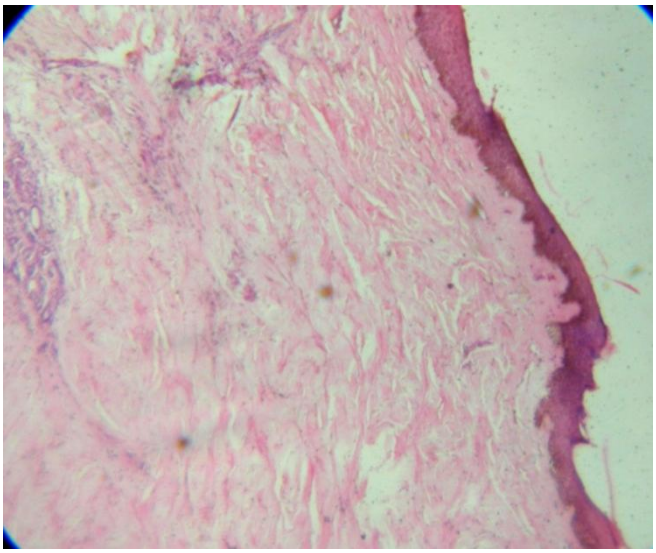
FIG 7.LINEAR VERRUCOUS NEVUS ASSOCIATED WITN PSORIASIS

LINEAR VERRUCOUS EPIDERMAL NEVUS



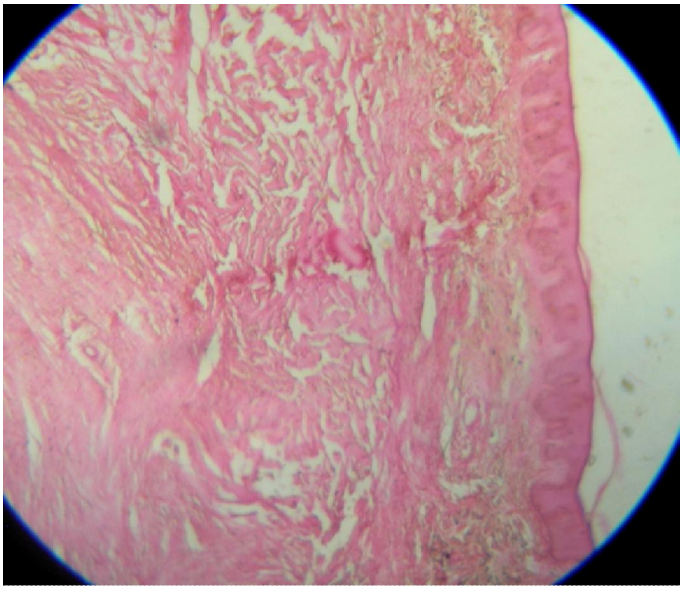
HPE OF LINEAR EPIDERMAL NEVUS SHOWING HYPERKERATOSIS, PAPILLOMATOSIS, ACANTHOSIS AND PATCHY INFILTRATES IN UPPER DERMIS

FIG 8.LINEAR MORPHOEA



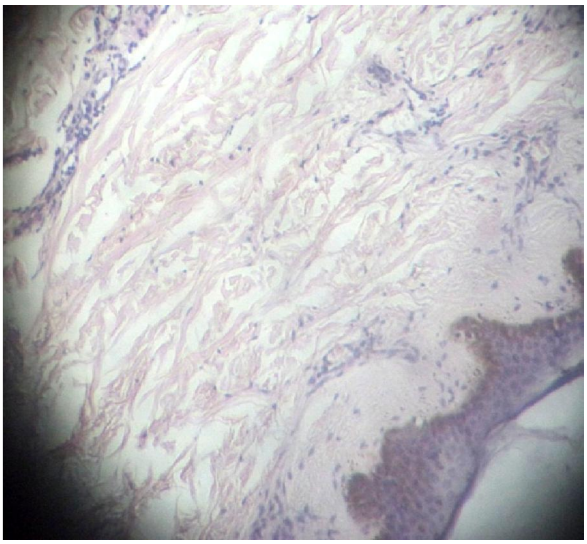
HPE OF LINEAR MORPHOEA SHOWING ATROPHIC EPIDERMIS,EOSINOPHILIC, OEDEMATOUS COLLOGEN BUNDLES IN UPPER DERMIS AND CUT SECTION OF ECCRINE DUCT

FIG 9.LINEAR MORPHOEA



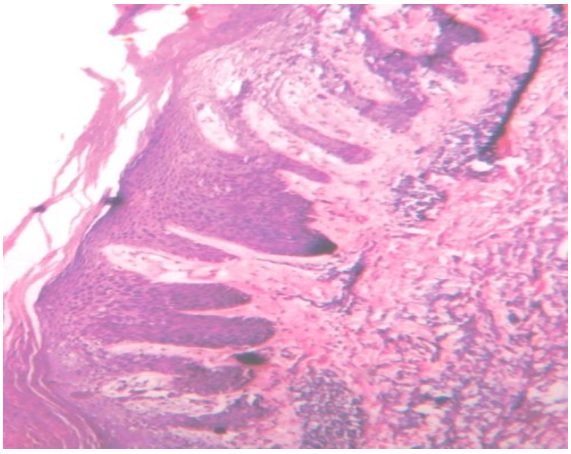
HPE OF LINEAR MORPHOEA SHOWING ATROPHIC EPIDERMIS, HOMOGENOUS THICKENED COLLAGEN BUNDLES AND HYPOCELLULAR DERMIS

FIG 10.PARRY ROMBERG SYNDROME



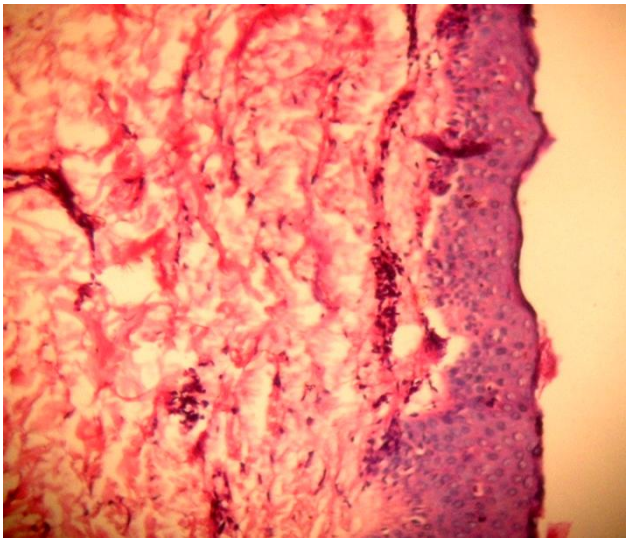
HPE OF PARRY-ROMBERG SYNDROME SHOWING ATROPHIC EPIDERMIS WITH HOMOGENOUS,HYPERTROPHIC COLLAGEN, HIGH UPTAKE OF ECCRINE GLANDS

FIG 11.LINEAR PSORIASIS



HPE OF LINEAR PSORIASIS SHOWING HYPERKERATOSIS, ABSENT GRANULAR LAYER, REGULAR ELONGATION OF RETE RIDGES

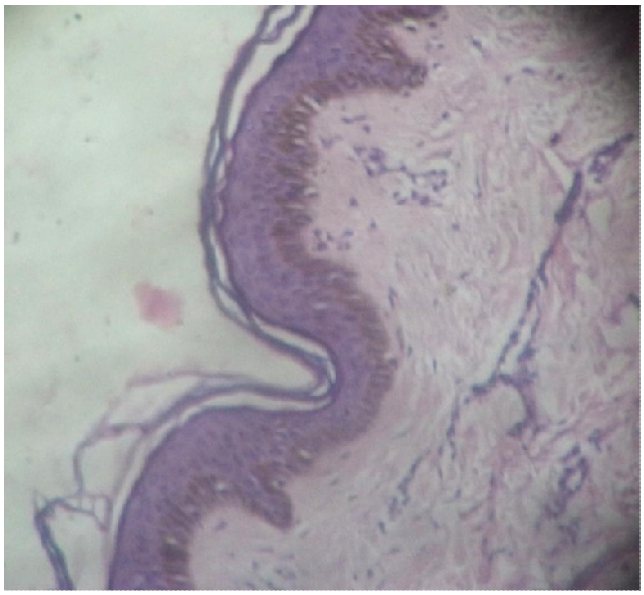
FIG 12.LINEAR VITILIGO



HPE OF LINEAR VITILIGO (TAKEN FROM THE FIRST PATIENT)- SHOWING ABSENCE OF MELANOCYTES AND LYMPHOCYTIC INFILTRATES IN DERMIS

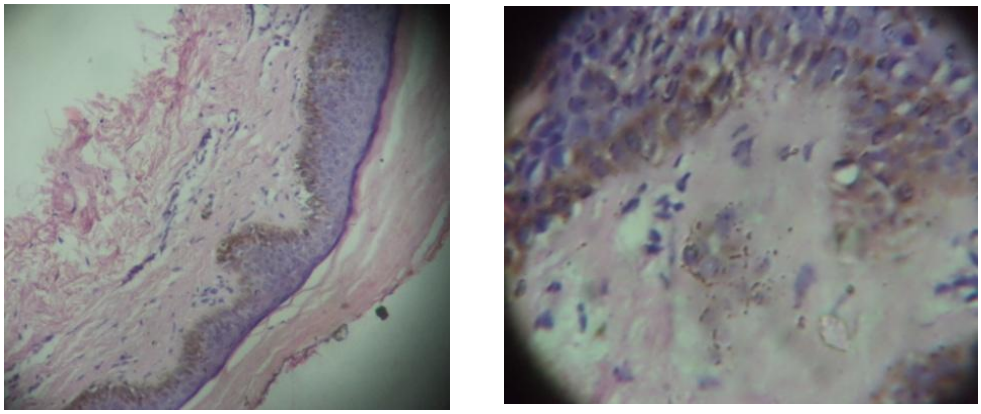
FIG 13. HYPOMELANOSIS OF ITO

FIG 14 . LINEAR AND WHORLED NEVOID HYPERMELANOSIS



HPE OF LWNH SHOWING INCREASED PIGMENTATION IN BASAL LAYER OF EPIDERMIS

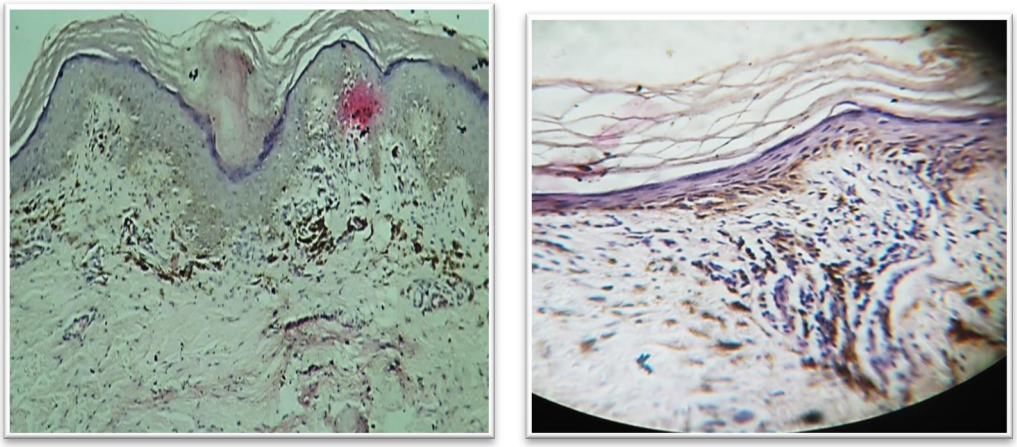
FIG 15. INCONTINENTIA PIGMENTI



LOW POWER VIEW **HIGH POWER VIEW**

HPE OF INCONTINENTIA PIGMENTI SHOWING INCREASED PIGMENT IN BASAL LAYER AND PIGMENT INCONTINENCE

FIG 16. LICHEN PLANUS PIGMENTOSUS



HPE OF LPP SHOWING
ATROPHIC EPIDERMIS,VACUOLAR DEGENERATION OF BASAL LAYER, PIGMENT INCONTINENCE, PLENTY OF MELANOPHAGES IN UPPER DERMIS
SPARSE LYMPHOHISTIOCYTIC INFILTRATE

DISCUSSION

In this study of 90 cases with a linear distribution, of the lesions which did not exhibit Koebner's phenomenon, none of the cases seemed to follow the linearity determined by the Nerves, vascular or lymphatic structure and it has been suggested that these lesions develop in the Lines of Blaschko.

Hence the various nevoid and acquired conditions which are supposed to follow the Lines of Blaschko, which are thought to be due to a form of human mosaicism were included in this study. Most of the patients were asymptomatic and mainly came for cosmetic reasons, except the Linear lichen planus patients and few of Lichen striatus patients who had presented in our Dermatology Outpatient Department for intense itching.

LICHEN STRIATUS

Lichen striatus formed the majority of cases amounting to 25 in this study. The condition is said to occur commonly in the age group of 5 - 15 years whereas in the present study, majority of patients were in the age group of between 3 – 20 years. It was more commonly observed in females in this study group with a Male : Female ratio 11:14 as also been documented by Hauber et al.[68]

The lesions are normally asymptomatic, with occasional pruritus in the study group as described in the literature. There were no predisposing factors in any of the patients, as Lichen striatus is of unknown etiology.

Most of the patients in the study group had lesions over the extremities, but few patients had lesions also over the trunk as recorded in the literature, which complied with the variable sites of expression. All the patients had unilateral distribution of the lesions. Two of the patients showed nail changes among this group, although changes in the form of longitudinal ridging, subungual hyperkeratosis, splitting and onycholysis have been documented.

Atopy was found to be associated with Lichen striatus in 80% of patients, although none in this study group had personal or family history of atopy but associated lesions like pityriasis veriscolor, xerosis and photosensitivity were seen, which were not documented so far and may be coincidental.

Histopathological examination showed a chronic dermatitis picture in majority of patients in this study group, few cases showed psoriasiform dermatitis like features and some other showed lichenoid dermatitis like picture which was consistent with the variable histological pictures as described in the literature. There were no systemic abnormalities noted in any of the Lichen striatus patients in this study group.

LINEAR LICHEN PLANUS

Linear lichen planus formed the next common condition in this study group consisting of 18 cases. Among them most of the patients were in the age group between 11 – 30 years and the average age at the time of diagnosis was 25 years. In this study Male: Female ratio of 11:7 was noted, showing a slight male preponderance in contrary to the literature.[69]

There was no history of contact with any chemicals or trauma but 2 patients had history of intake of NSAID and another 2 were incidentally associated with freckle and Becker's naevus.

There was no history of similar lesions in their family members or any other associated autoimmune disorders. It has been suggested that the linear distribution seen could be due to the tendency of Lichen Planus to develop, with the formation of clone of predisposed or vulnerable cells, which is predetermined during embryogenesis.

Most of the patients presented with the complaints of intense itching. These lesions started as hyperpigmented, discrete, flat topped papules with a violaceous hue distributed in a linear pattern. In some of the patients, the papules coalesced to form the linear plaques which were continuous or interrupted. Among 18 patients 2 patients had nail changes and one patient had mucous membrane involvement.

In sixteen patients, the lesions were found on the extremities. Two patients showed multiple linear lesions following the lines of Blaschko, but no immune compromised state was noted as shown in literature. The length of the lesions ranged from 5 cms - 50 cms and 4 patients had lesions extending the entire length of the limb.

On histopathological examination, sixteen out of 18 lesions showed the classical features of Lichen planus as described in the literature. [70] and one picture showed normal epidermis, basal cell degeneration, superficial mononuclear cell infiltrate in upper dermis, colloid bodies and pigment incontinence which was fit into the features of Lichenoid dermatitis. One case was diagnosed clinically as lichen planus but HPE showed features of Lichen planus pigmentosus (LPP).

LPP which usually manifests

- In contrast to lichen planus it is asymptomatic dark brown patches
- it has a longer clinical course
- Usually occurs in sun exposed site-face , neck and upper trunk
- Striking predominance of LPP in flexures -axilla and inguinal region

But in this case it has unusual presentation like

- Unilateral distribution
- linear pattern
- Lesions in unexposed areas

HbsAg seropositivity was found in one of the patient, which has not been reported in literature. Some of the associated features were Becker's nevus, pityriasis versicolor, Tinea corporis, Varicose vein, chronic urticaria , freckle which may be coincidental.

LINEAR EPIDERMAL NEVUS

In this study, Linear epidermal nevus accounted for 19 cases, with 13 cases between the age group of 1 and 20 years. Out of whom 12 were males and 7 were females. A male preponderance was seen in this study, in contrary to equal sex incidence given in the literature.[71] All the patients (19) with this disorder mainly came for cosmetic reason. No family history of similar lesion was recorded.

The lesions manifested as hyperpigmented verrucous papules arranged in a linear continuous or interrupted bands and majority of our patient the lesions were since birth, but in few of the patients the lesions became apparent later in life.

In this age group 5 patients had lesions involving trunk and extremities, implicating systemized form of verrucous epidermal nevus.

Most (17 out of 19) of the skin biopsies showed the classical features of verrucous epidermal nevus like hyperkeratosis, irregular acanthosis, papillomatosis, well formed granular layer, increased pigment basal layer and patchy inflammatory infiltrates in upper dermis. Two of them showed the features of Inflammatory Linear verrucous epidermal nevus like hyperkeratosis with foci parakeratosis, acanthosis, elongation and thickening of the rete ridges with a 'psoriasiform' appearance, papillomatosis, and spongiosis with exocytosis of lymphocytes.

LINEAR MORPHOEA

In this study nine cases of Linear Morphoea were recorded, among them 7 were less than 20 years of age. Generally the peak incidence of this condition is between 20 and 30 years of the age group.[72] Out of the 9 cases, 1 was male and 8 were female patients forming a Male: Female ratio 1:8 correlating with the female preponderance of this condition as recorded in the literature.[72]

Except one who had prior history of intramuscular injection over the lesional site, In all others(8) there was no history of any provocative factors like trauma or drug intake prior to the onset of lesion.

There was no history of similar lesion in the family members. Most of the patients with an asymptomatic atrophic plaques except one who had pain over the plaque. Most of the patients (4 out of 9) had lesions over the head and neck region among this one case was diagnosed as

Parry Romberg syndrome and 5 patients had lesions over the lower limb, which is the commonest site of involvement shown in literature.[73]

All the investigations pertaining to Morphoea were found to be normal including X– ray, except 4 patients who had eosinophilia and 3 cases that showed ANA positivity. Most(6 out of 9) of the skin biopsies showed the classical features of morphoea.

LINEAR PSORIASIS

In this study, among the two patient,one had presented with hypopigmented scaly plaque of 6 months duration over the extremity. There was no evidence of any trauma preceding the lesion. The patient had fine, regular pitting over the finger nails. On histopathological examination the lesion showed a characteristic feature of psoriasis, with which, the clinical diagnosis was revised from lichen striatus to linear psoriasis.

Second patient was presented with asymptomatic, erythematous plaques over the extremity on hisptopathological examination, it was found to have hypergranulosis and orthokeratosis, alternating with absent granular layer and parakeratosis and had other characteristic features of psoriasis.

SEGMENTAL VITILIGO

In this study group, three (2 female + 1 male) patients presented with segmental vitiligo between the age group of 8 years and 21 years which was earlier than the other types of vitiligo.

The lesions manifested as patches, arranged in a linear, continuous or interrupted bands, involving mainly the extremities. One had lesions over the face and neck region corresponding to the dermatomes rather than Blaschko's lines, perhaps in keeping with a neuronal etiological theory of vitiligo and it could be also clonal susceptibility of melanocytes to neurons.

One patient had leukotrichia over the vitiligo patches. There was no family history of similar lesions or any mucosal or nail involvement. None of them had vitiligo elsewhere in the body. There was no other associated auto immune disorders. Biopsy of these lesions showed the features consistent with vitiligo, like flaky hyperkeratosis, normal epidermis and absence of melanocytes in the basal cell layer and sparse inflammatory in upper dermis.

CONCLUSION

1. The Incidence of Linear Dermatoses in our Dermatology Out Patient Department, Govt. General Hospital, Madras Medical College, Chennai during the period of AUGUST- 2013 to JULY-2014 - 0.2 % .

2. Among the Linear Dermatoses, Lichen striatus was found to be more common.

3. The other Dermatoses following Blaschko's lines, in the descending order of frequency seen in this study were Linear epidermal Nevus, Linear Lichen Planus, Linear Morphoea, Inflammatory Linear Verrucous Epidermal Nevus, Segmental Vitiligo, Hypomelanosis of Ito, Linear lichenoid dermatitis, Linear psoriasis, Segmental Neurofibromatosis , Linear Whorled Nevoid Hypermelanosis, Incontinentia pigmenti, Nevus depigmentosus, Linear porokeratosis.

4. In this study, on the whole, slight female preponderance was noted.

5. Majority of patients showed unilateral distribution in a linear pattern, more often on the extremities.

6. The importance of histopathological correlation is very obvious. Cases which were clinically diagnosed as Lichen Striatus showed histopathological features of Psoriasis and Linear Epidermal

Verrucous Nevus. One case diagnosed clinically as Epidermal nevus was found to be superimposed with psoriasis histologically, in another case which was diagnosed as linear psoriasis clinically after histopathology the diagnosis was revised to linear porokeratosis. It shows the importance of histopathology which ultimately changing the management in any given condition.

7. The lesions were more of a cosmetic concern in most of the cases in this study.

8. Very few associations were noted in cases of Lichen Planus, Incontinentia Pigmenti, Epidermal nevus.

9. Cases of Lichen Planus were associated with Becker's nevus, Varicose vein, Freckles, HbsAg seropositivity, Chronic urticaria and Androgenic alopecia.

9. Incontinentia Pigmenti was associated with aplasia cutis, right limb hypotonia, limb length discrepancy, hypoplastic external genitalia and cicatricial alopecia.

10. In one patient of epidermal nevus was associated with ipsilateral gigantism.

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PATIENT CONSENT FORM

Title of the study : Study on linear dermatoses

Name of the Participant:

Name of the Principal(co-investigator): DR.P.SARASWATHY.

Name of the Institution: Department of Dermatology,

Rajiv Gandhi Government General Hospital, Chennai

Documentation of the informed consent

I _____ have read the information in this form(or it has been read for me).I was free to ask any questions and they have been answered. I am over 18 years of age and exercising my free power of choice, hereby give my consent to be included as a participant in the study.

1. I have read and understood this consent form and the information provided to me.
2. I have had the consent document explained to me.
3. I have been explained about the nature of the study.
4. My rights and responsibilities have been explained to me by the investigator.
5. I agree to cooperate with the investigator and I will inform him/her immediately if I suffer unusual symptoms.
6. I have not participated in any research study at any time.
7. I am aware of the fact that I can opt out of the study at any time without having to give any reason and this will not affect my future treatment in this hospital.

8. I hereby give permission to the investigators to release the information obtained from me as a result of participation in this study to the sponsors, regulatory authorities, Govt agencies and IEC.I understand that they are publicly presented.

9. My identity will be kept confidential if my data are publicly presented.

10. I am aware that if I have any question during this study, I should contact at one of the addresses listed above.

Participant's initials:_____

For adult participants:

Name and signature/thumb impression of the participant(or legal representative if participant incompetent)

_____ _____ _____

Name Signature Date

Name and signature of impartial witness(required for illiterate patients):

_____ _____ _____

Name Signature Date

Address and contact number of the impartial witness:

Name and Signature of the investigator or his representative obtaining consent:

_____ _____ _____

Name Signature Date

For Children being enrolled in research

Whether child's assent was asked : Yes No (Tick one)

[If the answer to the above question is Yes, write the following phrase:

You agree with the manner in which assent was asked for from your child and given by your child.

You agree to have your child take part in this study].

[If answer to the above question is No. give reasons (s) _____

Although Your Child did not or could not give his or her assent, you agree to your child's participation in this study].

Name and signature / thumb impression of the participant's parent(s) (or) legal representative):

(Name)_____ (Signature)_____ Date:

(Name)_____ (Signature)_____ Date:

Name and signature of impartial witness (required if parent of participant child illiterate):

(Name)_____ (Signature)_____ Date:

(Name)_____ (Signature)_____ Date:

Address and contact number of the impartial witness:_____

_____

Name and signature of the Investigator or his representative obtaining consent:

(Name)_____ (Signature)_____ Date:

(Name)_____ (Signature)_____ Date:

PROFORMA

Name : Date:

Age : O.P / I.P No.:

Sex : Informant (if Child):

Occupation :

Address :

HISTORY

Present History

Main Complaints :

Duration :

Distribution :

Progression :

Precipitating Factor: Drugs / Infection / Sun Exposure /

Stress / others _____

Past History :

Family History :

Treatment History :

Personal History :

Single / Married :

Consanguinity :

D.M / H.T / Smoking / Alcohol use / I.V Drug user

General Examination :

Built : Nourishment :

Pallor : Cyanosis :

Jaundice : lymphadenopathy :

Asso., Congenital Anomalies :

Systemic Examination :

Pulse rate: Blood Pressure :

CVS : RS :

CNS : P / A :

Eye : Skeletal :

Dental :

Dermatological Examination :

1. Basic Morphology :

Hyperpigmented / Hypopigmented

Scaly / Inflammed

Atrophic / Hypertrophic

Macule / papule / Nodule

Verrucous / Plaque / Vesicle

Comedones

Vascular

Hair Over the Lesion

Secondary Morphology:

Linear :

Topography :

1. Upper Limb
2. Lower Limb
3. Trumk
4. Head and Neck

Colour:

1. Hypopigmented
2. Skin Coloured
3. Blue
4. Black
5. Brown
6. Violaceous

Involvement of Underlying Structure Yes / No

Associated Skin Conditions

_____

Associated other Conditions

_____

Whether the Lesions following

Along the lines of BLASCHKO

Koebner's Phenomenon

Examination of

Hair : Nails :

Palms : Soles :

Mucous Membranes :

Investigations

Skin Biopsy No. :

Routine Inv : HB: TC :

DC :

ESR : RBS: Urine:

HBsAg : ANA : VDRL;

ELISA : LFT: RFT:

ECG : CXR: CT Scan:

Diagnosis:

Sn	M/f	Age	Consan	Past h/o	Family h/o	Syst- e/o	Colour	Distribu	Duration	Prog/st	Fixity	Ass-cut	Ass-oth	B/k	H,n,m	Biopsy	Inv/sp	Diag
21	F	9	3	NIL	NIL	N	2	2	2Y	p	-FIX	-	-	B	-	Y	ANA 1/40,E>	L.MOR
22	F	40	-	NIL	NIL	N	1	1	6M	p	-	-	-	B	-	Y	RBS	LS
23	F	1,1/2	-	NIL	NIL	N	5	2	1Y	p	-	-	-	B	-	-	N	LS
24	M	19	-	NIL	NIL	N	4	2	3Y	p	-	TV	-	B	NAIL INV	Y	N	LP
25	M	24	-	NIL	NIL	N	6	2	1Y	p	-	LP in elbow	-	B	-	Y	N	LP
26	F	70	-	simil l 10y back	NIL	N	5	2	2M	p	-	-	HT	B	-	Y	N	LP
27	F	13	-	NIL	NIL	N	1	3	8M	p	-	-	-	B	-	Y	N	ILVEN
28	F	13	-	NIL	NIL	N	1	2	1Y	s	-	-	-	B	-	-	N	LS
29	M	21	-	NIL	NIL	N	5	2	2M	p	-	-	-	B	PIT NAIL	Y	N	LP
30	F	44	-	NIL	NIL	N	5	1	3M	p	-	-	-	B	-	Y	N	ILVEN
31	F	8	-	NIL	NIL	N	1	4	3Y	s	-	-	-	B	-		N	SEG-VIT
32	F	7	-	NIL	NIL	N	4,5	1.2.3	6Y	p	-	U/L GIGAN	-	B	-	Y	N	LVEN
33	M	22	3	NIL	NIL	N	1	2	5Y	p	-	T-CR	-	B	-	Y	N	L.PORO
34	M	5	-	NIL	NIL	N	5	4	3Y	p	-	-	-	B	-	-	N	LVEN
35	F	45	-	NIL	NIL	N	6	2	8M	p	-	-	-	B	-	Y	RBS	LP
36	M	23	-	NIL	NIL	N	2	1	6M	p	FIX	WART	-	B	-	Y	ANA NEG,E>	L-MOR
37	F	41	3	NIL	NIL	N	6	2	2W	p	-	-	-	B	-	Y	N	LLD
38	F	11	2	NIL	NIL	N	5	1	3Y	p	-	-	-	B	-	-	N	LVEN
39	F	17	-	NIL	NIL	N	5	4	5Y	s	FIX	ALOPECIA	-	B	-	Y	ANA NEG.	L-MOR
40	F	15	-	NIL	NIL	N	1	1	7Y	s	-	-	-	B	-	Y	N	SEG-VIT
41	M	9	-	NIL	NIL	N	5	1	5Y	p	-	-	-	B	-	Y	N	ILVEN
42	M	27	-	NIL	NIL	N	4	1	4Y	p		ACROCHOR		B	-	Y	N	LVEN
43	M	52	3	NIL	NIL	N	6	2	8M	p		-	-	B	-	Y	N	LP

Sn	M/f	Age	Consan	Past h/o	Family h/o	Syst- e/o	Colour	Distribu	Duration	Prog/st	Fixity	Ass-cut	Ass-oth	B/k	H,n,m	Biopsy	Inv/sp	Diag
1	M	18	-	NIL	NIL	N	5	4	10Y	p	-	-	-	B	-	Y	N	LVEN
2	F	8	-	NIL	NIL	N	1	2	1Y	S	-	-	-	B	-	-	N	LS
3	F	42	-	NIL	NIL	N	1	2	8M	P	-	-	-	B	-	Y	N	LS
4	M	23	-	NIL	NIL	N	6	2	6M	P	-	-	-	B	-	Y	HBS-Ag +	LP
5	F	55	-	NIL	NIL	N	5	1	10Y	s	-	LSC	-	B	-	Y	N	LVEN
6	F	7	3	NIL	NIL	N	4	1	6Y	P	-	-	-	B	-	-	N	LVEN
7	F	22	-	NIL	NIL	N	1	2	4M	P	-	TV	-	B	-	Y	N	LS
8	M	29	-	NIL	NIL	N	1	3	4Y	s	-	-	CSOM	B	-	Y	N	LS
9	M	14	-	NIL	NIL	N	6	1	2Y	p	-	TC	-	B	-	Y	N	LP
10	M	42	-	NIL	NIL	N	1	2	25Y	s	-	TBVC	-	B	-	y	MAN-NEG	LVEN
11	M	28	-	NIL	NIL	N	5	1	2Y	p	-	SCALP PSORA	-	B	PITTING	Y	N	L.PSORA
12	M	11	-	NIL	NIL	N	1	4	3M	p	-	IBA	-	B	-	-	N	LS
13	F	11M	-	NIL	NIL	N	4,5	2	4M	p	-	-	-	B	-	-	N	LS
14	M	20	-	NIL	NIL	N	4,6	4	2M	p	-	-	-	B	-	Y	N	LP
15	M	29	-	NIL	NIL	N	1	2,4	2Y	s	-	-	-	B	-	Y	N	LS
16	M	22	-	NIL	NIL	N	1	1	1Y	s	-	TC	-	B	nail-split	Y	N	LS
17	M	3M	2	NIL	NIL	N	1	2	2M	p	-	-	-	B	-	-	N	HM OF ITO
18	F	10	2	NIL	NIL	N	1,5	1,2,3	10y	s	-	-	-	B	C.ALOPECIA	Y	N	IP
19	F	13		NIL	NIL	N	1	2	2Y	s	-	IBA	-	B	-		N	LS
20	M	17		NIL	NIL	N	2	4	5Y	p	-	-	-	B	-	Y	N	LVEN

Sn	M/f	Age	Consan	Past h/o	Family h/o	Syst- e/o	Colour	Distribu	Duration	Prog/st	Fixity	Ass-cut	Ass-oth	B/k	H,n,m	Biopsy	Inv/sp	Diag
44	M	21	-	NIL	NIL	N	1,5	4	15Y	p	-	-	-	B	-	Y	N	LVEN/PSO
45	M	7	-	NIL	NIL	N	1	4	3Y	s	-	leucotrichia	-	B	-	-	N	SEG-VIT
46	F	15	-	i.m,inj	NIL	N	6	2	1Y	p	FIX	ECZEMA	-	B	-	Y	ANA NEG,	L-MOR
47	F	16	-	NIL	NIL	N	5	2	1Y	p	-	-	-	B	-	Y	N	LP
48	M	8	3	NIL	NIL	N	1	3	6M	p	-	-	-	B	-	-	N	LS
49	F	27	-	NIL	NIL	N	6	1	2Y	p	-	-	-	B	-	Y	N	LP
50	M	18	-	NIL	NIL	N	5	4	18Y	p	-	-	-	B	-	Y	N	LVEN
51	M	54	-	NIL	NIL	N	6	2	1M	p	-	VARICOSE	-	B	-	Y	N	LP
52	F	17	-	NIL	NIL	N	5	4	10Y	p	-	-	-	B	-	Y	M	LVEN
53	F	34	-	NIL	NIL	N	6	1	6M	p	-	nail pits	-	B	-	Y	N	L-PSORA
54	M	50	2	NIL	NIL	N	2	3	30Y	s	-	PEM-VUL	DM	B	-	-	BS-PP	SEG-NF
55	M	40	-	NIL	NIL	N	1	4	39Y	s	-	-	-	B	-	-	N	N-DEPIG
56	F	42	-	NIL	NIL	N	5	2	20Y	s	-	-	-	B	-	Y	N	LVEN
57	F	30	-	NIL	NIL	N	1	2	1Y	p	-	-	-	B	-	Y	N	LS
58	F	40	-	NIL	NIL	N	1	3	9M	p	-	-	-	B	-	Y	N	LS
59	F	12	3	NIL	NIL	N	5	2	10M	p	-	-	-	B	-		N	LS
60	M	14	-	NIL	NIL	N	1	1	8M	P	-	-	-	B	-	Y	N	LS
61	F	44	-	NIL	NIL	N	5	1	44Y	S	-	-	-	B	-	Y	N	ILVEN
62	F	3	-	NIL	NIL	N	5,2	2,4	2Y	p	FIX		-	B	-	Y	EARLY,ANA NEG	L-MOR
63	M	22	-	NIL	NIL	N	4	2,3	10Y	p	-	ACNE	-	B	-	Y	LIPID PROFILE	LVEN
64	F	24	-	NIL	NIL	N	5	2	7Y	p	-	FRECKLE	-	B	ORAL LP	Y	N	LP
65	F	45	3	NIL	NIL	N	2	1	8M	p	-	SRS	-	B	-	Y	N	LS
66	M	19	-	NIL	NIL	N	1	2	6M	p	-	-	-	B	-	Y	N	LS
67	F	19	-	NIL	NIL	N	3,2	4	2Y	p	FIX	-	-	B	-	Y	ANA-1/10,E>	L-MOR
68	F	36	-	NIL	NIL	N	6	2	6M	p		CH-URTICARI	-	B	-	Y	N	LP

Sn	M/f	Age	Consan	Past h/o	Family h/o	Syst- e/o	Colour	Distribu	Duration	Prog/st	Fixity	Ass-cut	Ass-oth	B/k	H,n,m	Biopsy	Inv/sp	Diag
69	M	13	-	NIL	NIL	N	5	4	10Y	p	-	-	-	B	-	-	N	LVEN
70	M	15	-	NIL	NIL	N	4	4	10Y	p	-	SCABIES	-	B	-	-	N	LVEN
71	M	10	-	NIL	NIL	N	1	2	4Y	s	-	-	-	B	-	Y	N	LS
72	M	3	3	NIL	NIL	N	5	2	5M	p	-	-	-	B	-	-	N	LS
73	F	9	-	NIL	NIL	N	5	2,3	3M	p	FIX	ALOPECIA		B	-	Y	MRI,ANA NEG	L-MOR
74	F	5	-	NIL	NIL	N	2	4	4M	p	-	-	-	B	-	-	N	LS
75	M	2	-	NIL	NIL	N	1	2,3	6M	p	-	-	-	B	-	-	N	LS
76	M	14	-	NIL	NIL	N	5	4	2Y	p	-	PHRYNO	-	B	-	Y	N	LVEN
77	F	15	-	NIL	NIL	N	1	1,3	2Y	p	-	-	-	B	NAIL	Y	N	LS
78	M	17	-	NIL	NIL	N	1	1	7M	p	-	SRS	-	B	-	-	N	LS
79	F	4	-	NIL	NIL	N	1,2	1,2	3M	p	FIX	ALOPECIA	BELLS-P	B	-	Y	ANA1/40, E>	L-MOR
80	F	22	-	NIL	NIL	N	2	4	2Y	s	FIX	-	D-MASTI	B	-	Y	MRI,CT,X -RAY	PAR-ROM
81	M	43	-	NIL	NIL	N	4	2	1Y	p	-	-	-	B	AGA	Y	N	LP
82	F	42	-	NIL	NIL	N	2	2	2Y	p	-	-	-	B	-	Y	N	LP
83	M	15	-	NIL	NIL	N	4	4	5M	p	-	BECKERS	-	B	-	Y	N	LP
84	M	14	-	NIL	NIL	N	4,5	3,2,1	9Y	s	-		-	B	-	Y	MRI-ECHO-CT-N	LWNH
85	M	13	-	NIL	NIL	N	5	4	2Y	p	-	INTERTRIGO	-	B	-	Y	N	LVEN
86	M	7	-	NIL	NIL	N	6	2	8M	p	-	-	-	B	-	Y	N	LP
87	F	10	-	NIL	NIL	N	5	1,2,3	8Y	p	-	-	-	B	-	Y	N	LVEN
88	M	24	-	NIL	NIL	N	6	2	3M	p	-	FDE	-	B	-	Y	N	LLD
89	F	3	-	NIL	NIL	N	I	3,2	3Y	s	-	--	-	B	-		N	HM OF ITO
90	F	14	-	NIL	NIL	N	1	3,2	13Y	s	-	PRP	EPILEPSY	B	-	Y	MRI,CT-N	HM OF ITO

KEY TO THE MASTER CHART

S.N - serial number

M - male

F - female

Consan - consanguinity

N - normal

Colour code-1-hypo pigmented

2-skin coloured

3-blue

4-black

5-brown

6-violaceous

Distribution-1-upper limb

2-lower limb

3-trunk

4-head and neck region

Duration- w- weak

m-month

y -years

p- prograssive

s- static

Y- biopsy done

ass-cut- associated cutaeous lesions

ass-oth-associated other system lesion

B- Along the Blaschko's lines

H- Hair

N- Nail

T- Teeth

MM- Mucous membrane

CT- CT-scan

MRI-magnetic resonance imaging

BS-blood sugar

DM-diabetis mellitus

IBA-insect bite allergy

TV-tinea versicolor

FIX-fixed

MAN-mantoux

FDE-fixed drug eruption

PEM-VUL-pemphigus vulgaris

D-MASTI- difficulty in mastigation

E>- Increased eosinophilia

LS - Lichen striatus

LP - Lichen planus

LVEN - Linear verrucous epidermal nevus

LLD - Linear Lichenoid dermatitis

L.Psora- Linear psoriasis

ILVEN- Inflammatory Linear verrucous epidermal nevus

L.mor -Linear morphoea

HM of ITO- Hypomelanosis of ITO

PAR-ROM- Parry Romberg Syndrome

SEG-VIT- Segmental vitiligo

LWNH- Linear Whorled Nevoid Hypermelanosis

SEG-NF- Segmental Neurofibromatosis

IP- Incontinentia pigmenti

L.PORO- Linear Porokeratosis

ABBREVIATIONS

CHILD	-	Congenital hemidysplasia with ichthyosiform nevus and limb defects
MIDAS	-	*mi*crophthalmia, *d*ermal *a*plasia, *s*clerocornea
PEDDON	-	Porokeratoticeccrineostial and dermal duct nevus
GVHD	-	Graft Versus Host Disease
ILVEN	-	Inflammatory Linear verrucous epidermal nevus
CMI	-	Cell mediated immunity
HI	-	Hypomelanosis of ITO
BCIE	-	Bullous congenital ichthyosiform erythroderma
LVEN	-	Linear verrucous epidermal nevus
LLD	-	Linear Lichenoid dermatitis
LWNH	-	Linear and Whorled Nevoid Hypermelanosis
IP	-	Incontinentia pigmenti
PPK	-	Palmo Plantar Keratoderma
BCC	-	Basal cell carcinoma
EMF	-	Erythema multiforme
BLAISE	-	Blaschkos line associated inflammatory skin eruptions
ANA	-	Anti nuclear antibody
USG	-	Ultra sono gram
AD	-	Autosomal dominant
CNS	-	Central nervous system
HIV	-	Human immuno deficiency virus

INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI -3

EC Reg. No. ECR/270/Inst./TN/2013
Telephone No. : 044 25305301
Fax : 044 25363970

CERTIFICATE OF APPROVAL

To
Dr. P.Saraswathy,
PG in M.D. Dermatology, Venerology, Leprology
Department of Dermatology,
Madras Medical College, Chennai -3.

Dear Dr.P.Saraswathy,

The Institutional Ethics Committee of Madras Medical College reviewed and discussed your application for approval of the proposal entitled **"STUDY ON LINEAR DERMATOSES"**

No. 14102013

The following members of Ethics Committee were present in the meeting held on 08.10.2013 conducted at Madras Medical College, Chennai -3.

1. Dr. G. Sivakumar, MS FICS FAIS -- Chairperson
2. Prof. R. Nandini, MD -- Member Secretary
   Director, Instt.of Pharmacology, MMC, Ch-3 -- Member
3. Prof. Ramadevi,
   Director i/c, Instt.of Biochemistry, Chennai.
4. Prof. P. Karkuzhali, MD -- Member
   Professor, Instt.of Pathology, MMC, Ch -3.
5. Prof. KalaiSelvi. MD -- Member
   Prof. of Pharmacology, MMC, Ch -3.
6. Thiru. S. Govindasamy, BA BL -- Lawyer
7. Tmt. Arnold Saulina, MA MSW -- Social Scientist

We approve the proposal to be conducted in its present form.

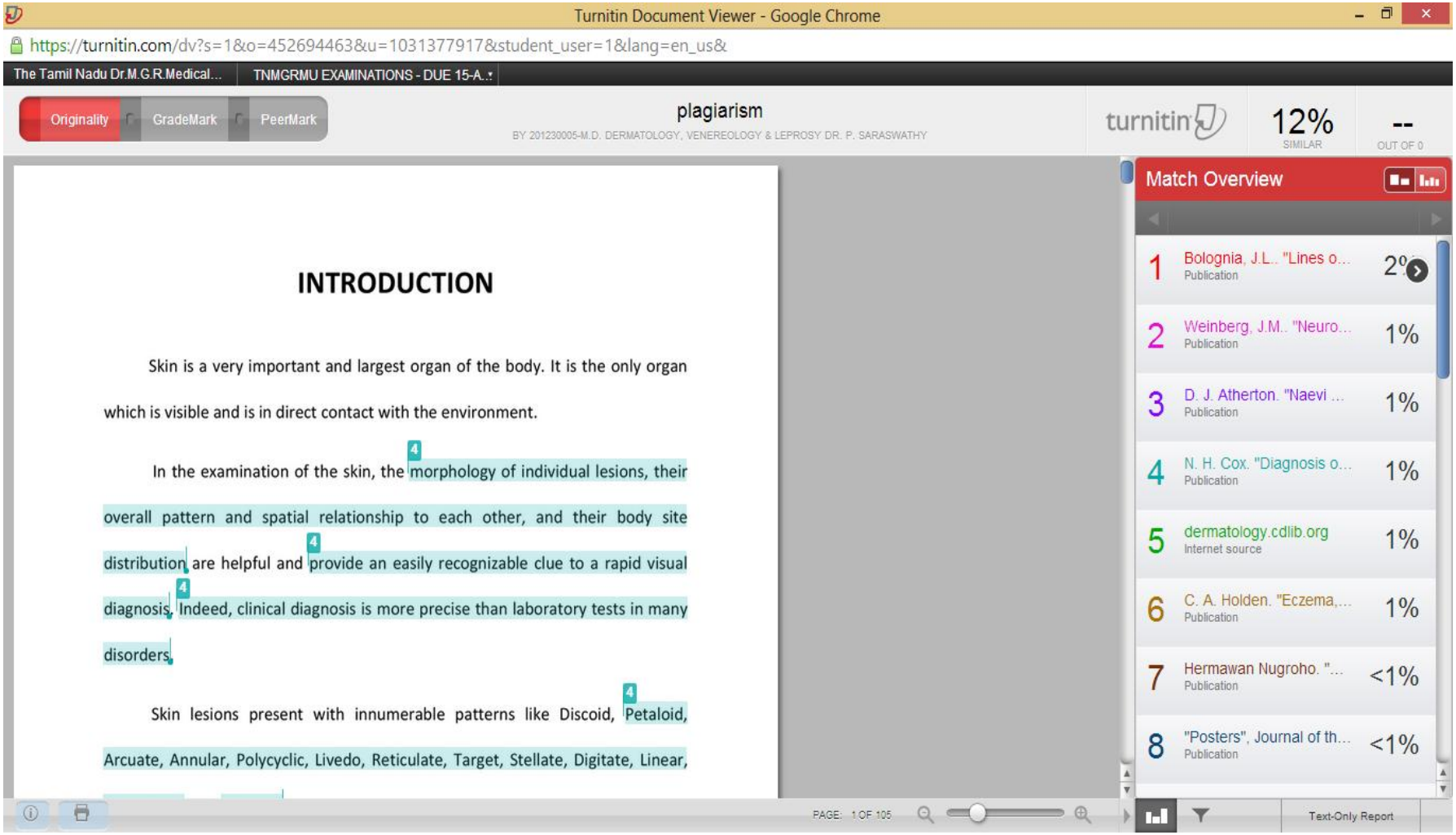
Sd/ Chairman & Other members

The Institutional Ethics Committee expects to be informed about the progress of the study, and SAE occurring in the course of the study, any changes in protocol and patients information / informed consent and asks to be provided a copy of the final report.

18/10/13

Member Secretary, Ethics Committee

18.07.14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INTRODUCTION
Skin is a very important and largest organ of the body. It is the only organ which is visible and is in direct contact with the environment.
In the examination of the skin, the morphology of individual lesions, their overall pattern and spatial relationship to each other, and their body site distribution are helpful and provide an easily recognizable clue to a rapid visual diagnosis. Indeed, clinical diagnosis is more precise than laboratory tests in many disorders.
Skin lesions present with innumerable patterns like Discoid, Petaloid, Arcuate, Annular, Polycyclic, Livedo, Reticulate, Target, Stellate, Digitate, Linear,
PAGE: 1 OF 105
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